5th Edition

Textbook of OPHTHALMOLOGY

HV Nema • Nitin Nema

DVD Contents

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Textbook of Ophthalmology

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5th Edition

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To
Pratibha and Sandeep

Preface to the Fifth Edition

More than twenty years have passed since the textbook of ophthalmology was first published. During the last twenty years, ophthalmology has undergone a phenomenal growth. Successive editions of the textbook have reflected some of the advances made in ophthalmic sciences. In the present fifth edition, all chapters have been updated, and revised or rewritten. The general format of the book is not changed although the clinical presentation has become more colorful. Almost every chapter is liberally illustrated with colored clinical photographs and drawings. Additional material is added to cover newer clinical entities, diagnostic procedures and therapeutic or surgical modalities of treatment of eye diseases. However, no attempt has been made to present an exhaustive text and the book has retained its original virtues of accuracy, clarity, comprehensiveness and consistency of style.

An appendix is included in this edition of the book. It contains drawing, description and uses of ophthalmic instruments. Surgical steps of common eye surgeries are demonstrated with the help of video clips. Observation of these video clips will certainly help students to understand and learn basic operative procedures. It will indeed enhance the interest of students in fascinating ophthalmic surgery.

At the end of each chapter, a few important references are given to enable the inquisitive student to gather more information on a particular topic of interest.

Although the book is written mainly for the undergraduate medical students to grasp the basic ophthalmology, hopefully, it will be beneficial for the postgraduate students and residents in ophthalmology. The book will be useful for the practicing general ophthalmologists in their day-to-day care of patients.

HV Nema
Nitin Nema

Preface to the First Edition

In spite of introduction of modern audiovisual techniques: projection slides and television tapes, the bedrock of undergraduate education in ophthalmology remains the standard textbooks. The need for a new and updated textbook is strongly felt with rapid developments and advancements in the ophthalmic sciences.

A thorough knowledge of ocular anatomy and physiology is essential for a proper understanding of ophthalmology. Hence, general anatomy and physiology of the eye and its individual structures are comprehensively dealt with.

The reader may find the chapter on *Examination of the Eye* informative and interesting as all the routine advanced clinical methods with diagnostic techniques in ophthalmology are incorporated in brevity.

To bring out a better understanding of ocular therapeutics, a separate chapter is included. Common diseases of the eye have been described in some detail and their salient clinical features are highlighted with the help of black and white and coloured photographs. Recent advances in the treatment of ophthalmic disorders have also been covered. The chapter on *Operations upon the Eyeball and its Adnexa* presents important surgical techniques in present day use. The emergence of community ophthalmology signifies a shift from curative to preventive approach in the speciality.

The extensive use of tables and illustrations has been made in an effort to focus the attention of readers on easy differential diagnosis and better retention of facts.

The present textbook is primarily written for undergraduate medical students. However, it is hoped that general practitioners and diploma students of ophthalmology would also find it useful in revision and updating of their basic knowledge on the subject.

HV Nema

Acknowledgements

To write a textbook of this kind requires the cooperation and help from colleagues, students, friends and publishers. It is gratifying to mention that we received the help in plenty from all corners. The source of each illustration is acknowledged throughout the pages of the book. However, we would like to express our sincere indebtedness to following colleagues for their generosity in providing a large number of excellent colored photographs and/or video clips from their archives at a short notice.

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2. Professor YR Sharma, MD, Dr RP Center for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi
3. Professor Anita Panda, MD, Dr RP Center for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi
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6. Dr Jotirmay Biswas, MS, Head, Uveitis and Pathology Department, Sankara Nethralaya, Chennai
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We are indeed grateful to M/S Appasamy Associates, Chennai for permitting us to reproduce excellent drawings of the ophthalmic instruments from their atlas.

We record our sincere thanks to Professor MK Singh, MS, Eye Department, BHU, Varanasi for his assistance in the preparation of early editions of the book. Mr Tapan Chaurasia, MSc and Mr Lalit Gupta deserve our special thanks for editing colored photographs and video of ophthalmic surgical procedures, respectively.

Last but not the least, the credit of meticulous publication of this textbook of ophthalmology goes to Shri Jitendar P Vij, Chairman and Managing Director, and Mr Tarun Duneja, General Manager (Publishing) Jaypee Brothers Medical Publishers (P) Ltd., New Delhi, and their enthusiastic staff.

HV Nema
Nitin Nema

Contents

CHAPTER

1 Anatomy of the Eyeball

ANATOMY

The eyeball (Fig. 1.1) lies in a quadrilateral pyramid-shaped bony cavity situated on either side of the root of the nose called *orbit*. Each eyeball is suspended by extraocular muscles and their fascial sheaths. There lies a pad of fat behind the eyeball to provide a protective cushion.

At birth the eyeball measures antero-posteriorly about 17.5 mm and reaches 24 mm in adults. The horizontal and vertical diameters of the eyeball are 23.5 mm and 23.0 mm, respectively. As it is flattened from above downwards its shape resembles with an oblate spheroid.

The central points on maximum convexities of the anterior and posterior curvatures of the eyeball are called *anterior* and *posterior poles* (Fig. 1.2). The axis of the eyeball passes through the poles. The line joining the poles is called *meridian*. The optic nerve leaves the eyeball 3 mm medial to the posterior pole and passes along the axis of the orbit, therefore, the axes of the eyeball and the orbit do not coincide but make an angle between them.

The eyeball is composed of three concentric tunics (Fig. 1.3).

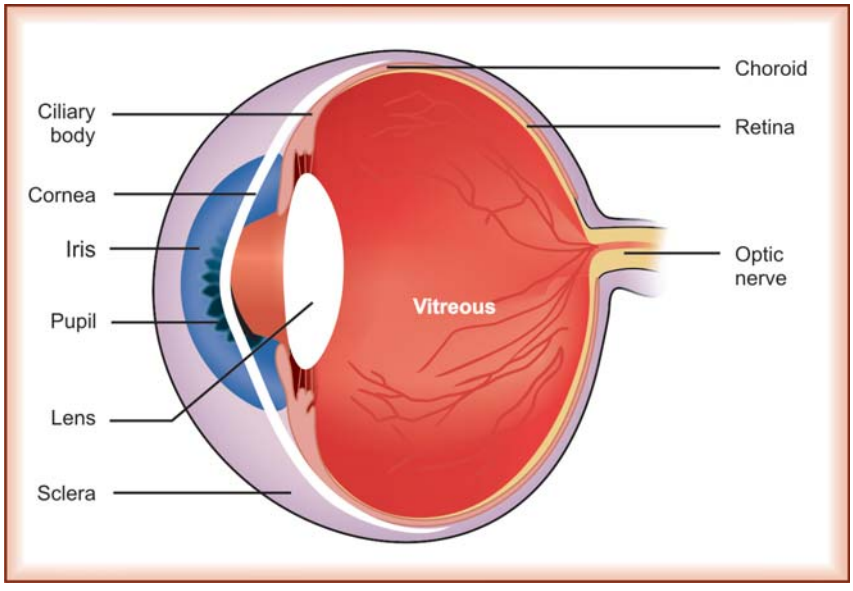

Fig. 1.1: A sagittal section through the eyeball

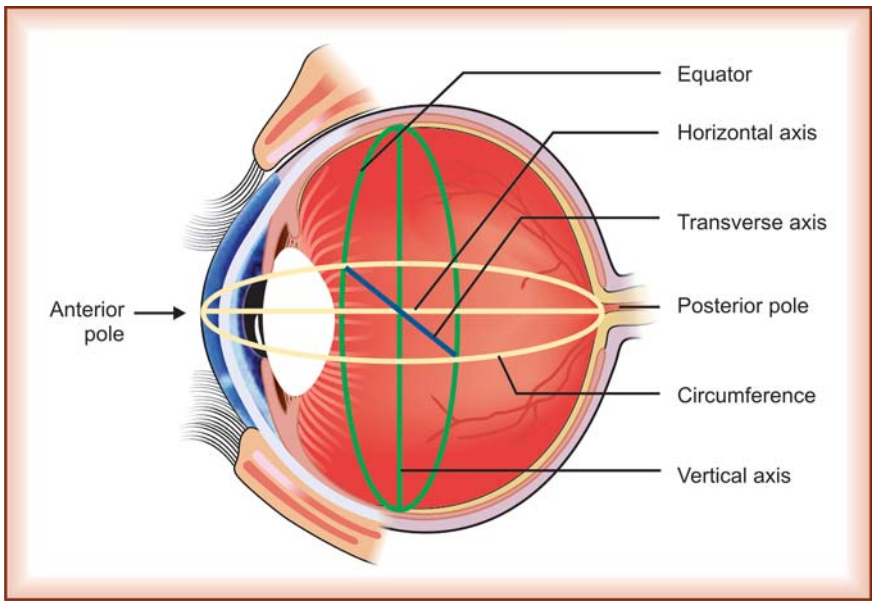

Fig. 1.2: The poles, axes, meridians and equator of the eyeball

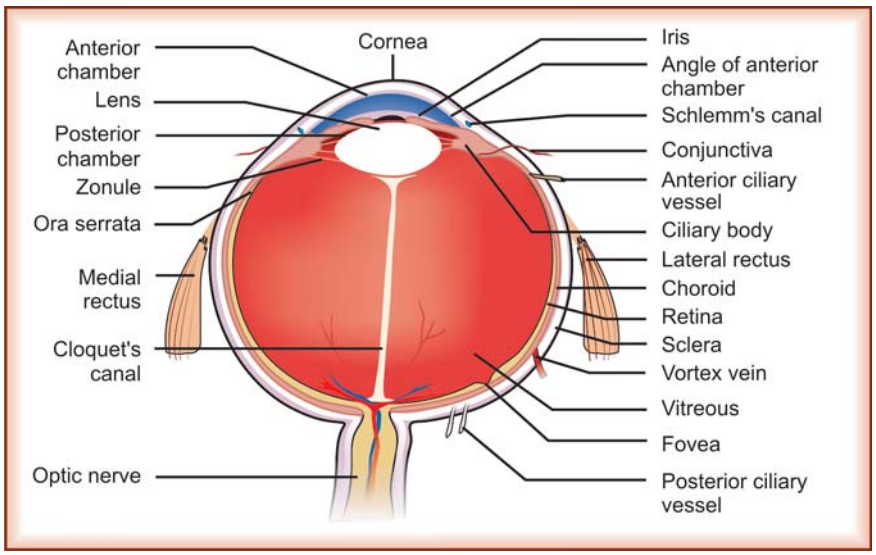

Fig. 1.3: Diagram of horizontal section of eyeball

The *outer tunic* consists of anterior one-sixth transparent part, the *cornea*, and the remainder five-sixths opaque part—the *sclera*.

The *intermediate vascular tunic* comprises from behind forward—the *choroid*, the *ciliary body* and the *iris*.

The *innermost sentient layer*, the *retina*, serving the primary purpose of photoreception and transformation of light stimuli, is connected with the central nervous system by a tract of nerve fiber, the *optic nerve*.

The anterior part of the sclera is covered by a mucous membrane, the *conjunctiva*, which is reflected over the lids and also adhered firmly around the periphery of the cornea—the *limbus*.

The eyeball can be divided into an anterior and a posterior segment.

The *anterior segment* consists of the cornea, anterior chamber, iris, posterior chamber, lens and ciliary body.

The *posterior segment* is formed by the vitreous cavity filled by vitreous humour, retina, choroid and optic nerve.

The lens is suspended from the ciliary body by fine delicate fibrils called *suspensory ligament of the lens (zonule)*.

The *anterior chamber* is bounded anteriorly by the posterior surface of the cornea and posteriorly by the anterior surface of the iris and the lens. It has a peripheral recess known as the *angle of the anterior chamber* through which the drainage of aqueous humor takes place.

The *posterior chamber* is lined anteriorly by the posterior surface of the iris and posteriorly by the ciliary body and the zonule. Both the chambers contain aqueous humor and communicate with each other through the pupil.

Blood Supply of Eyeball

The arteries of the eyeball are derived from the *ophthalmic artery*, a branch of *internal carotid artery*. The retina gets its blood supply from the central retinal artery, a branch of ophthalmic artery, which enters the optic nerve about 10 mm behind the eyeball. The *central retinal artery* gives pial branches in the intraorbital and the intravaginal course. After running outward and forward it reaches the optic nerve head and gives superior and inferior papillary branches, from each of which come off a nasal and a temporal branch. Each branch continues to divide dichotomously spreading over the retina and reaching the ora serrata.

The veins of retina do not accurately follow the course of the arteries, except at the disk, where they join to form the central retinal vein which accompanies the central retinal artery.

The uveal tract is supplied by *ciliary arteries* arranged into three groups—the *short posterior*, the *long posterior* and the *anterior ciliary arteries* (Fig. 1.4).

The *short posterior ciliary arteries* (20 in number) pierce the sclera around the optic nerve and supply the choroid.

The *long posterior ciliary arteries* (2 in number) pierce the sclera obliquely in the horizontal meridian on either side of the optic nerve and run anteriorly between the sclera and the choroid without giving off any branch. They divide in the ciliary body and anastomose with the anterior ciliary arteries to form *circulus arteriosus major* at the root of iris.

The *anterior ciliary arteries* are derived from the muscular branches of the ophthalmic artery to the four rectus muscles. They pierce the sclera 3 to 4 mm behind the limbus to join the long posterior ciliary artery. Before piercing they give off branches to the conjunctiva, limbus and episclera.

The *venous drainage of the uveal tract* occurs through the *ciliary veins* which form three groups—the *short posterior ciliary*, the *venae vorticosae* and the *anterior ciliary*. The short posterior ciliary veins receive blood only from the sclera, and the anterior ciliary veins from the outer part of the ciliary muscles. The bulk of the blood is drained through the venae vorticosae (*vortex veins)* comprising four large trunks which open into the ophthalmic vein.

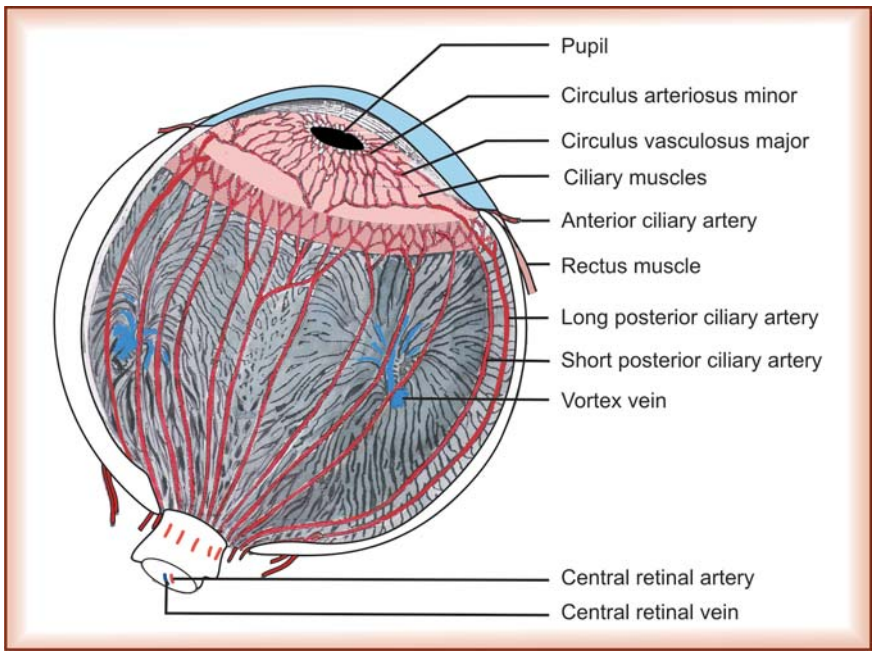

Fig.1.4: Blood supply of eyeball

Nerve Supply of Eyeball

The sensory nerve supply to the eyeball is derived from the *ophthalmic division of trigeminal* nerve (Fig. 1.5).

It comes mainly by the nasociliary nerve either directly through the long *ciliary nerve* or indirectly through the short ciliary nerves.

The long ciliary nerves (2 in number) pierce the sclera in the horizontal meridian on either side of the optic nerve and run forwards between the sclera and the choroid to supply the iris, ciliary body, dilator pupillae and cornea.

The *short ciliary nerves* (about 10 in number) come from the *ciliary ganglion* and run a wavy course along with the short ciliary arteries. They give branches to the optic nerve and ophthalmic artery and pierce the sclera around the optic nerve. They run anteriorly between the choroid and the sclera, reach the ciliary muscles where they form a plexus which innervates the iris, ciliary body and cornea.

The *motor root of ciliary ganglion*, derived from the branch of *oculomotor nerve* to inferior oblique, supplies the sphincter pupillae and ciliary muscles.

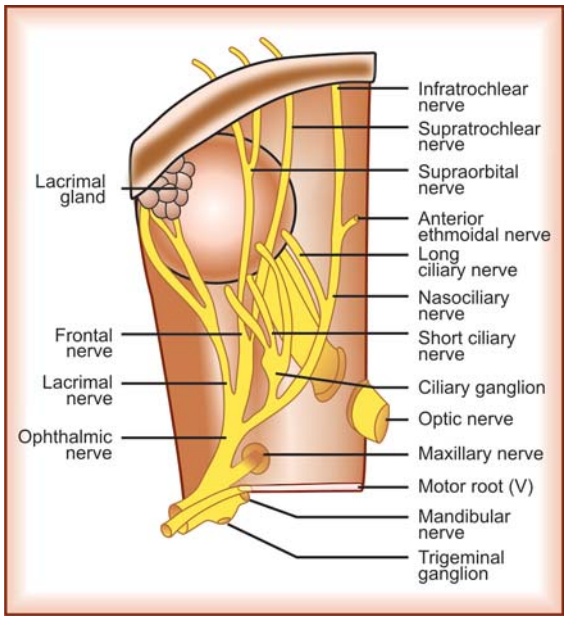

Fig. 1.5: Nerve supply of eyeball

DEVELOPMENT OF THE EYE

The eyeball develops as a part of the central nervous system. The latter develops from the neural groove which invaginates to form the neural tube. A thickening appears on either side of the anterior part of the tube which grows at 4 mm human embryo stage to form the *primary optic vesicle* (Fig. 1.6).

The vesicle comes in contact with the surface ectoderm and invaginates to form the *optic cup.* The inner layer of the cup forms the future retina, epithelium of ciliary body and iris, and sphincter and dilator pupillae, while the outer layer forms a single layer of pigment epithelium. At the anterior border of the cup paraxial mesoderm invades to form the stroma of the ciliary body and the iris.

Development of Lens

The development of the lens begins early in embryogenesis. When optic vesicles enlarge they come in contact with surface ectoderm.

Lens plate: The surface ectoderm overlying optic vesicle thickens at about 27 days of gestation and forms the lens plate or lens placode (Fig. 1.7).

Lens pit: A small indentation appears in the lens plate at 29th day of gestation to form the lens pit which deepens and invaginate by cellular multiplication.

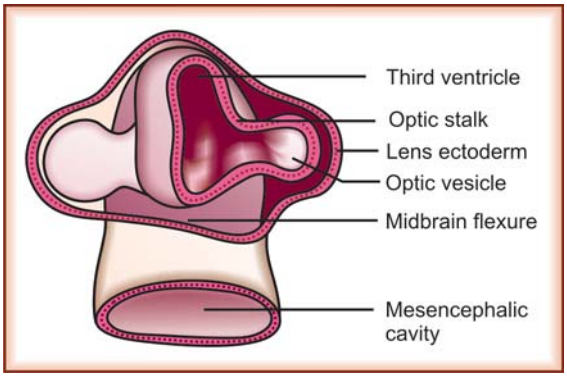

Fig. 1.6: Forebrain and optic vesicle in a 4 mm human embryo

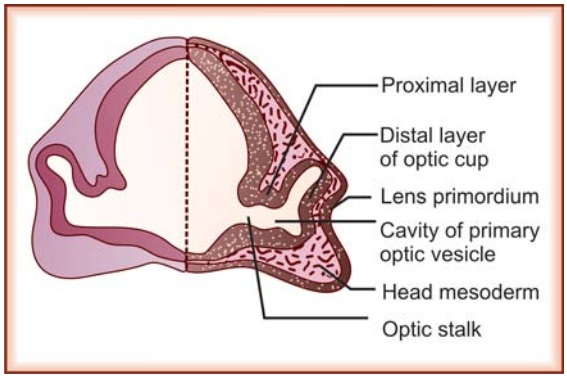

Fig. 1.7: Transverse section through forebrain of a 5 mm human embryo

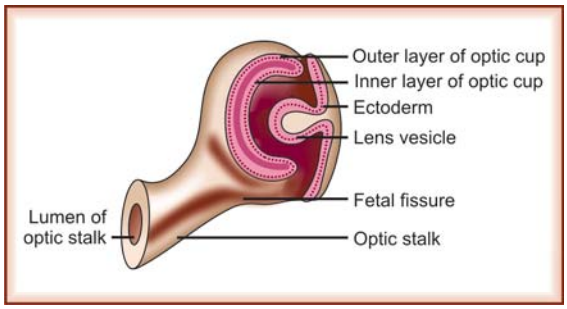

Fig. 1.8: Formation of lens vesicle

Lens vesicle: At about 33 days of gestation, lens vesicle (Fig. 1.8) is formed due to continued invagination of the lens pit, which later detaches from the surface ectoderm. The lens vesicle is a single layer of cuboidal cells that is encased within a basement membrane, the lens capsule.

Primary lens fibers: At about 40 days of gestation, the posterior cells of lens vesicle elongate to form the primary lens fibers. They fill up the cavity of the lens vesicle and form the embryonic nucleus.

Secondary lens fibers: The cuboidal cells of the anterior lens vesicle, also known as the *lens epithelium*, multiply and elongate to form the secondary lens fibers. The fibers formed between 2 and 8 months of gestation form the fetal nucleus.

Optic Stalk and Optic Fissure

A deep groove appears on the ventral surface of the optic cup and stalk, called *fetal fissure*. Through the optic fissure mesenchyme enters the optic cup in which the hyaloid system of vessels develop to provide nourishment to the developing lens (Fig. 1.9).

The vascular system gradually atrophies with the closure of the optic fissure and is replaced by the vitreous, presumed to be secreted by the surrounding neuroectoderm. The hollow optic stalk is filled by the axons of ganglion cells of the retina forming the optic nerve. The condensation of the mesoderm around the optic cup differentiates to form the outer coats of the eyeball (choroid and sclera) and structures of the orbit.

The stroma of the cornea, the anterior layer of the iris and the angle of the anterior chamber are formed by the mesodermal condensation, while the corneal and the conjunctival epithelium develop from the surface ectoderm.

A cleft is formed due to the disappearance of the mesoderm lying between the developing iris and cornea, the anterior chamber (Fig. 1.10). The canal of Schlemm appears as a vascular channel at about fourth month of gestation.

Ocular Adnexa

The eyelids develop from both the surface ectoderm and the mesoderm. The medial and lateral parts of the frontonasal process join to form the upper lid, while the maxillary process forms the lower lid. Cilia develop from the epithelial buds. The ingrowth of inferior canaliculus cuts off a portion of the lid forming the lacrimal caruncle.

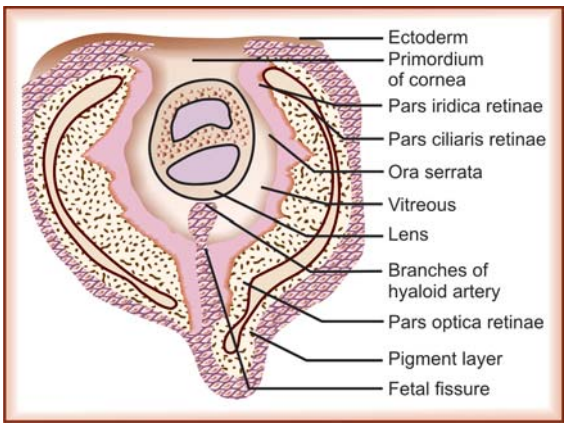

Fig. 1.9: Diagram showing eye and fetal fissure of a 15 mm human embryo

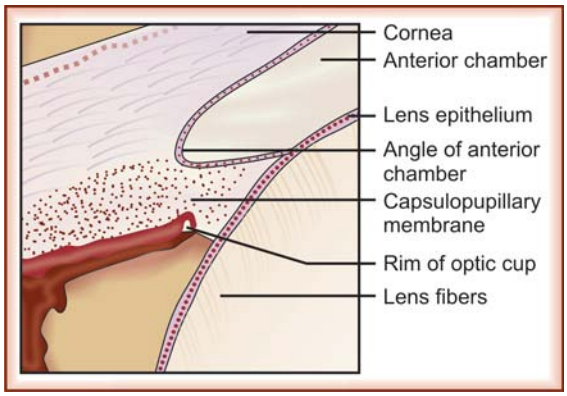

Fig. 1.10: Development of angle of anterior chamber at 75 mm human embryo stage

Eight epithelial buds from the superolateral part of the conjunctiva form the lacrimal gland. A solid column of cells from the surface ectoderm form the primordium of lacrimal sac. The growth of ectoderm upward into the lid and downward into the nose forms canaliculi and nasolacrimal duct, respectively. The canalization of the cellular columns starts at about third month and is completed by seventh month of intrauterine life.

The extraocular muscles develop from preotic myotomes which are innervated by the oculomotor, trochlear and abducent nerves. The individual extraocular muscle differentiates at about 20 mm stage of developing embryo excepting the levator palpebrae superioris which develops from the superior rectus at a later stage.

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CHAPTER

2 Physiology of the Eye

AQUEOUS HUMOR

The eye is a peripheral organ of vision. It subserves its function due to the optically transparent media, particularly the cornea and the lens, which focus the images of the objects on a sensitive layer—the retina. The eye maintains its shape by intraocular pressure. The avascular structures, lens and cornea, receive their nourishment by aqueous humor. The formation and circulation of aqueous humor and the maintenance of intraocular pressure are important aspects of physiology of the eye.

Aqueous humor is a clear fluid, filling the anterior and the posterior chambers of the eye. Its refractive index is 1.336 and viscosity 1.025 to 1.040. The osmotic pressure of aqueous humor is slightly higher than plasma. The aqueous contains glucose, urea, proteins, inorganic salts, ascorbic acid, lactic acid and some dissolved oxygen.

The walls of capillaries of iris and ciliary body, two layers of ciliary epithelium and walls of retinal capillaries constitute a system of semipermeable membranes, separating blood from the ocular cavity, known as *blood-aqueous barrier.* This barrier is relatively impermeable so that the large-sized molecules from plasma cannot pass into the eye. Such a mechanism is necessary for the maintenance of optical transparency of aqueous humor. However, breakdown of blood-aqueous barrier, as a result of ocular trauma or inflammation, permits the escape of whole blood or its turbid proteinous contents into the aqueous—*plasmoid aqueous*.

Formation of Aqueous Humor

For several years aqueous humor was considered as a *simple filtrate from blood*. Owing to a significant difference in the chemical compositions of aqueous and blood such a concept was rejected. Subsequently a *theory of ultrafiltration* from the capillaries of ciliary processes was postulated; but it could not explain all the facts regarding the higher concentration of ascorbates. Therefore, a hypothesis of *active ciliary secretory process* was proposed. It has been found that the rate of transport of sodium by the ciliary epithelium is sufficient to explain the rate at which water enters the ocular cavity. The active transport of sodium by the ciliary epithelium is carried out by a sodium pump demonstrated by Berggren and Cone. The energy required for the active transport to a large extent is provided by citric acid cycle. The active process of aqueous formation is a complex series of biochemical reactions wherein the role of many enzymes and their linkage with Na^+- K^+ ATPase yet remain unknown. A combination of ultrafiltration (25%) and active ciliary secretory process (75%) explains the formation of aqueous humor.

Circulation of Aqueous Humor

The circulation of aqueous humor is essential for regulation of the intraocular pressure as well as for metabolic activities of the intraocular structures. Aqueous humor is drained out by two routes: (i) trabecular meshwork route and (ii) uveoscleral route (Fig. 2.1).

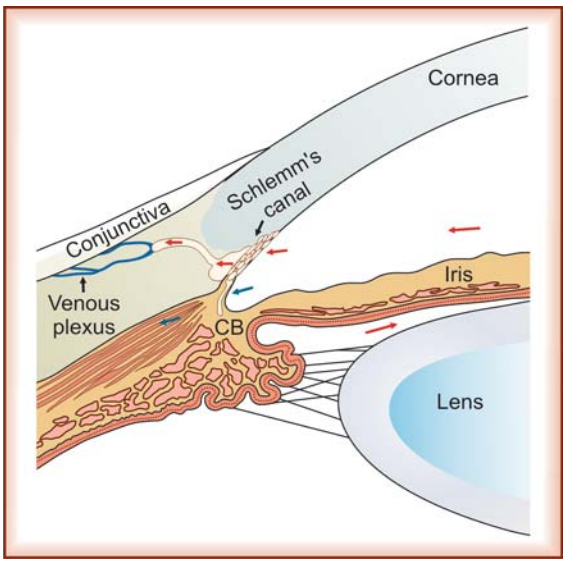

Fig. 2.1: Drainage of aqueous humor

Trabecular Outflow

The trabecular route accounts for bulk of aqueous outflow (75 to 90%). The formed aqueous humor is collected in the posterior chamber, flows through the pupil into the anterior chamber and finally escapes through the drainage channels at the angle of the anterior chamber. The aqueous filters through trabecular meshwork at the angle of the anterior chamber into the canal of Schlemm and from there a number of aqueous veins and efferent channels drain it into the episcleral veins and intrascleral venous plexus, respectively. Approximately 2 µl (1% of the fluid in the anterior chamber) of the aqueous humor drains away per minute. The drainage of aqueous humor is influenced by the patency of exit channels and the venous pressure just within the sclera. Improper cleavage of the angle of the anterior chamber causes a rise in the intraocular pressure often seen in congenital glaucoma. If the venous pressure is raised the drainage of aqueous humor is embarrassed which can be demonstrated on episcleral vein in the *glass rod phenomenon experiment*. The episcleral vein at the site of junction with the aqueous vein presents a laminated appearance due to blood and aqueous running side by side. When the vein is compressed by a glass rod, fluid flows from the vessel with higher pressure into the one with lower. If the venous pressure is higher there is blood influx into the aqueous vein, if the pressure in the aqueous vein is higher, aqueous influx is seen in the vein.

Uveoscleral Outflow

Nearly 10-26% of aqueous outflow occurs through the uveoscleral route. The aqueous passes across the ciliary body into the suprachoroidal space and is drained in the venous circulation.

Intraocular Pressure

Intraocular pressure (IOP) is the pressure inside the eyeball. It is determined by the rate of aqueous production by the ciliary epithelium and the amount of its drainage through the trabecular meshwork. The gradient of pressure in the ocular capillaries across which the fluid transfer takes place greatly influences the intraocular pressure. Normally there is a balance between the rate of formation of the aqueous humor and its drainage, hence, wide ranging fluctuations do not occur. The intraocular pressure in a human eye usually ranges from 12 to 20 mm Hg. It is most accurately measured by a manometer. However, manometric measurements are not possible in human eyes and, therefore, a measurement of the degree of

indentation of cornea by a standard weight is utilized. Such a method is called *indentation tonometry* (Schiotz). The variation in the rigidity of sclera induces significant error in such measurements warranting correction. A more dependable method, *applanation tonometry*, is in wide clinical use. The measurements are usually expressed as mm Hg and the intraocular pressure is referred as *ocular tension*. The normal mean ocular tension is 15.4 + 2.5 mm Hg by applanation and 16.1 + 2.8 mm Hg by Schiotz tonometer. Usually the intraocular pressure does not vary significantly between the two eyes. A consistent difference of 4 to 6 mm Hg between the two eyes is known as *Downey's sign* and is an indication for investigation for glaucoma.

What factors regulate the intraocular pressure at its normal level are not known clearly. However, there is evidence to suggest that a center in the hypothalamus exercises a control on the intraocular pressure to maintain its homeostatic equilibrium. The afferent path from the eye to this center is not determined, but the efferent path runs through the sympathetics traveling down the spinal cord relaying in the cervical ganglion and reaching the eye by way of the cervical chain and the ophthalmic artery. Probably, the center is also responsible for the diurnal variations of intraocular pressure which is often seen. The average variation throughout the day is about 2 mm Hg. A rise may occur in the morning hours which is mainly due to the changes in the rate of aqueous humor production.

Following factors tend to alter the intraocular pressure.

1. *Variation in the Hydrostatic Pressure in the Capillaries*
   A rise in the ciliary capillary pressure often results in a rise in intraocular pressure and *vice versa*. Vasodilatation does not lead to an increased pressure, a low pressure is a frequent outcome.
2. *Variation in the Osmotic Pressure of the Blood*
   A change in the osmotic pressure of the blood alters the process of diffusion across the capillary wall; hypotonicity induces a rise and hypertonicity a fall in intraocular pressure.
3. *An Increase in the Permeability of the Capillaries*
   An increased permeability of the ciliary capillaries results in the formation of plasmoid aqueous which induces a rise in the osmotic pressure of the aqueous and thus increases the intraocular pressure. The IOP further accentuates if particulate materials block the drainage channels.
4. *Change in the Volume of the Eyeball*
   Generally, a small volumetric change in the eyeball is normally compensated by an increased drainage mechanism. However, big tumors, intraocular hemorrhages and sudden vasodilatation induce pressure changes due to indistensibility of the sclera.
5. *Obstruction in the Circulation of Aqueous*
   Blockage at the pupil and/or at the angle of the anterior chamber results in profound rise in intraocular pressure.
6. *Alteration in Aqueous Formation*
   Alteration in the secretory activity of the ciliary epithelium should hypothetically alter the intraocular pressure. Hypersecretion of the aqueous causes a rise and hyposecretion a fall in intraocular pressure.
   Besides these, changes in the pH of blood, topical and systemic drugs, general anesthesia and psychological stress are known to alter the intraocular pressure.

METABOLISM OF OCULAR TISSUES

The ocular tissues consist of both vascularized and nonvascularized structures. Iris, ciliary body, choroid and retina do not differ in general metabolic activity from other tissues of the body. The retina has a very high metabolic rate and it rapidly dies if its blood supply is cut off even for a

short time. The nonvascularized structures of the eye such as cornea and lens derive their energy requirements from phosphorylation of carbohydrates and auto-oxidative system.

Cornea requires energy for the maintenance of its transparency. There are possibly three important routes for the transport of metabolites to and from the cornea—perilimbal capillaries, aqueous humor and tears. Glucose enters the cornea either by simple diffusion or by active transport through aqueous humor. The atmospheric oxygen presumably dissolved in tears enters the epithelium. The oxygen may also reach the cornea through tear film from the palpebral conjunctival vessels especially when eyelids are closed. It reaches the deeper layers of the cornea through aqueous humor. The breakdown of glucose occurs by aerobic and anaerobic processes into carbon dioxide and water, and lactic acid, respectively.

The lens derives its energy mainly through carbohydrates. Its metabolism is a complex one that has not been fully understood properly. The lens, a structure devoid of blood vessels, has low metabolic rate as the rates of consumption of oxygen and utilization of glucose are far lower than many other ocular tissues. The amino acids and fatty acids are oxidized in the mitochondria of lens epithelium via citric acid cycle. The carbohydrate metabolism in the lens can be described under the following heads.

Glycolysis

Glycolysis is an anaerobic process wherein glucose is phosphorylated and subsequently broken down to pyruvic acid to form lactic acid with the help of a number of enzymes. This is the main route of metabolism of glucose in the lens. About 80% of glucose is matabolized through glycolysis.

Citric Acid Cycle

Citric acid cycle is an oxidative metabolic process occurring in the mitochondria. As there is a paucity of oxidative enzymes and mitochondria in the lens, the Krebs' cycle is very inactive. The lens derives oxygen from the aqueous. The glucose metabolism through Krebs' citric acid cycle produces CO_2 and H_2O as its end products and provides 38 molecules of ATP. It generates about 20% of the total ATP from glucose in the lens. The metabolic activity of lens is mostly confined to the lens cortex. The central nucleus is more or less inert.

Hexose Monophosphate (HMP) Shunt or Pentose Phosphate Pathway

In this shunt glucose is phosphorylated and then oxidized through co-enzyme triphosphopyridine nucleotide (TPN); the oxidative decarboxylation occurs with the production of carbon dioxide and phosphorylated pentoses (ribose-5 phosphate). The latter is a constituent of nucleotides in DNA/RNA and in coenzymes. The HMP shunt is very active in the lens.

Sorbitol Pathway

The sorbitol pathway is not of much significance because nearly 5% of the glucose used by the lens is metabolized by this pathway. However, it assumes greater significance in sugar cataract.

PHYSIOLOGY OF VISION

When light falls upon the retina, two essential changes, photochemical and electrical, occur. The photochemical changes occur in the pigments of the rods and cones. The light breaks the rod pigments, rhodopsin which is a chromoprotein, into yellow retinene (aldehyde of vitamin A) and, eventually, to colorless vitamin A. The reaction is reversible. This photochemical reaction initiates the visual response and induces changes in the electrical potential which are transmitted through the bipolar cells to the ganglion cells and then along the fibers of the optic nerve to the brain. The electrical changes vary in frequency with the intensity of light and can be recorded by electroretinogram (ERG).

The stimulation of the retina with light yields three types of sensations—light sense, form sense and color sense.

Light sense is the faculty which permits us to perceive light of all gradation of intensities. The minimum amount of light energy which can induce a visual sensation is known as *light minimum*. The light minimum is very small if the eye is dark-adapted and it increases when the rods and cones are diseased. After ascertaining the light minimum, if the intensity of light is gradually increased one can appreciate a difference in the amount of illumination, called *light difference*. The least perceptible difference of illumination bears a constant relation to the total illumination and is known as *Weber's law*. The light difference is also influenced by the adaptation of the eye and is increased in disorders affecting the optic nerve.

Form sense is the faculty which enables us to perceive the shape of the objects in the outer world. It is the function of cones, and is most acute at the fovea where cones are packed densely. The ability to distinguish the shapes of the objects is called *visual acuity* or *central vision*. Besides retinal, the form sense is mostly psychological. It includes the light sense, the sense of position and the sense of discrimination.

Color sense is the faculty by which eye distinguishs different colors and color tones. There are three primary colors—red, green and blue. The cones are responsible for the recognition of colors. Colors are better appreciated in day light while in dim light they look gray (Purkinje's shift).

The cones contain three types of photopigments which absorb red, green and blue wavelengths of light. The 3 different spectral classes of cones are short-wavelength sensitive (*S-cones*), middle-wavelength sensitive (*M-cones*) and long-wavelength sensitive (*L-cones*). The spectral sensitivity of S-cones peak at around 440 nm, M-cones peak at 545 nm and L-cones peak at 565 nm, but they may have overlapping sensitivities as well. The presence of three types of photoreceptors in the retina helps in perceiving the colors (*Young-Helmholtz trichromatic theory of color vision*). Each color receptor responds to all wavelengths (long-red, middle-green and short-blue). Admixture of these primary colors can produce other colors of the spectrum. The trichomatic theory has limitations in explaining color confusion and complementary color after-images.

Hering proposed an *opponent color theory* which suggests that there are three sets of receptor systems: red-green, blue-yellow, and black-white. The stimulation or excitation of one results in inhibition of the opposite receptor in the pair. This concept can explain color contrast and color blindness to some extent.

The most widely accepted theory of color vision is *stage theory* that incorporates both the trichomatic theory and the opponent color theory into 2 stages. The first stage is the receptor stage consisting of 3 photopigments. The second is the neural processing stage where color opponency occurs at the post-receptoral level.

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CHAPTER

3 Neurology of Vision

VISUAL PATHWAY

The visual sensations are perceived by the rods and cones and conducted to the brain through three sets of neurons. The conducting nerve cells or neurons of the first order are the bipolar cells of the inner nuclear layer of the retina with their axons in the inner plexiform layer. The neurons of the second order are the ganglion cells, the axons of which pass in the nerve fiber layer and along the optic nerve to the lateral geniculate body. From here the neurons of the third order transmit the impulse through the optic radiations to the visual center situated in the occipital lobe.

The arrangement of the nerve fibers in the optic nerve is peculiar; the fibers from the peripheral parts enter the periphery of the optic nerve, while the fibers from the adjoining parts of the disk enter the central part of the nerve. The macular fibers or papillomacular fibers initially enter the nerve on the outer aspect, but they soon become more centrally arranged in the posterior part of the nerve. These fibers undergo a partial decussation in the chiasma wherein the nasal fibers cross, while the temporal ones enter the optic tract of same side. Similarly, the fibers of the peripheral retina form two separate groups. If a vertical line is drawn through the macula, it divides the retina into two halves—temporal and nasal. The fibers from the nasal half enter the chiasma, decussate and pass into the opposite optic tract. The fibers from the temporal half enter the chiasma and pass into the optic tract of same side. Both uncrossed and crossed fibers pass to alternating laminae in the lateral geniculate body. The neurons of the third order pass in the optic radiations to reach the occipital lobe.

A lesion of an optic tract or of one occipital lobe leads to blindness of temporal half of the retina on the same side and of the nasal half of the retina on the opposite side. In other words it causes loss of vision in the opposite half of the binocular field of vision, a defect known as *homonymous hemianopia*. The visual fibers in optic radiations are closely related to the internal capsule, temporal lobe of the brain and the lateral ventricle. The fibers are liable to be compressed in cases of tumors of the temporal lobe and distention of the lateral ventricle.

The visual center is situated in and about the calcarine fissure of the occipital cortex. The part above the calcarine fissure represents the upper corresponding quadrants of both retinas and the part below, the lower quadrants. The posterior part of the occipital lobe represents the macula.

Pupil

The central opening in the iris is called *pupil*. The pupillary size varies between 1 and 8 mm. It tends to be smaller in newborn (parasympathetic tone) and elderly persons (decreased sympathetic

activity). Pupils are wider in teenagers and in dim light. They are relatively dilated in the state of joy, fear or surprise due to increased sympathetic tone. The average diameter of pupil in an adult is 4 mm in ordinary room light. The pupil during sleep is usually constricted due to reduced tone of dilator pupillae and diminution of inhibitory impulses to the constrictor center. Abnormal constriction of pupil is called *miosis*, while abnormal dilatation is known as *mydriasis*.

The main function of pupil is optical. Constriction of pupil regulates the entry of light inside the eye and allows the retina to adapt to the changes in the illumination. Constriction of pupil cuts off the peripheral and chromatic aberration and astigmatism, it also increases the depth of perception.

PUPILLARY PATHWAYS AND REACTIONS

The pupil is controlled by two muscles—sphincter and dilator pupillae. The former constricts the pupil while the latter dilates. The sphincter pupillae is supplied by the cholinergic nerves of parasympathetic system through the III cranial nerve (Fig. 3.1), while the dilator by the adrenergic fibers of the cervical sympathetic nerves (Fig. 3.2).

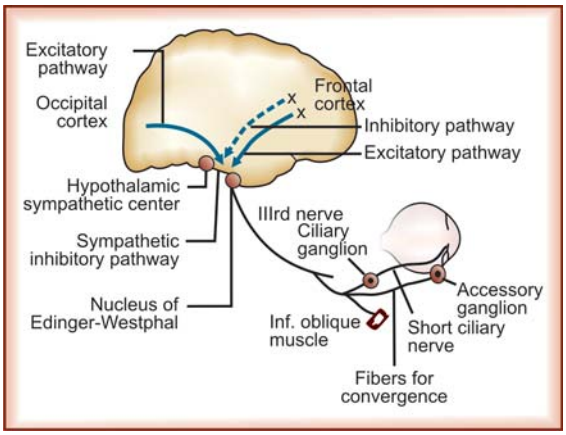

Fig. 3.1: Parasympathetic pupillary system

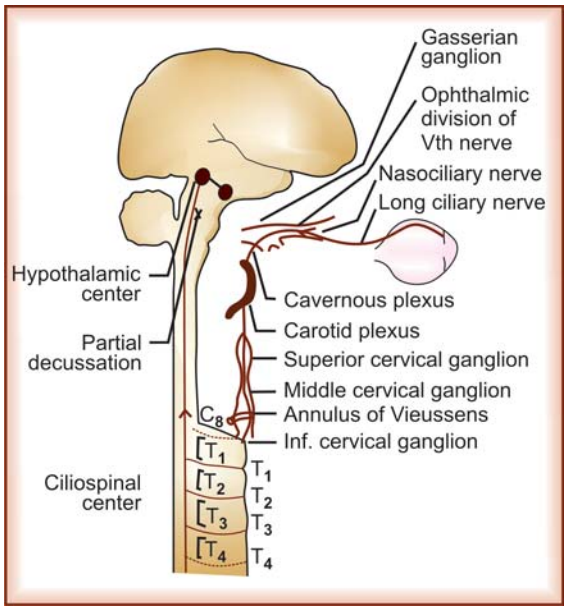

Fig. 3.2: Sympathetic pupillary system

The pupils take part in the following three reflexes which are of clinical importance.

1. Light reflexes
2. Near reflex, and
3. Psychosensory reflex.

When light above a threshold value enters the eye the pupil constricts, which is called *direct light reflex*, while the constriction of the contralateral pupil is known as *consensual light reflex*.

Light Reflex

The afferent pathways of light reflex follow the course of visual fibers (optic nerve) and undergo semi-decussation in the chiasma. The pupillary fibers travel along the visual fibers in the optic tract. Just before reaching the lateral geniculate body they leave the tract to enter the superior collicullus and the pretectum. The pupillary fibers are relayed in the pretectal nucleus. The neurons from pretectum, after getting partially decussated in the midbrain, project their axons to Edinger-Westphal group of oculomotor nucleus, and thus

distribute the impulses of each tract to both III nerve nuclei. This decussation is important because it is responsible for the consensual light reflex (Fig. 3.3).

The efferent pathways of the sphincter mechanism is controlled by Edinger-Westphal nucleus which lies under the aqueduct of Sylvius. The efferent impulses run through the III nerve to the ciliary ganglion lying in the muscle cone. The postganglionic fibers relay through the short ciliary nerves to innervate the sphincter pupillae.

Near Reflex

The constriction of pupil occurs on looking at a near object or with convergence and accommodation. Basically, near reflex is not a true reflex but an associated reaction.

Convergence Reflex

The constriction of pupil in near reflex is independent of any change in illumination. It is initiated by the fibers from the medial rectus muscles which

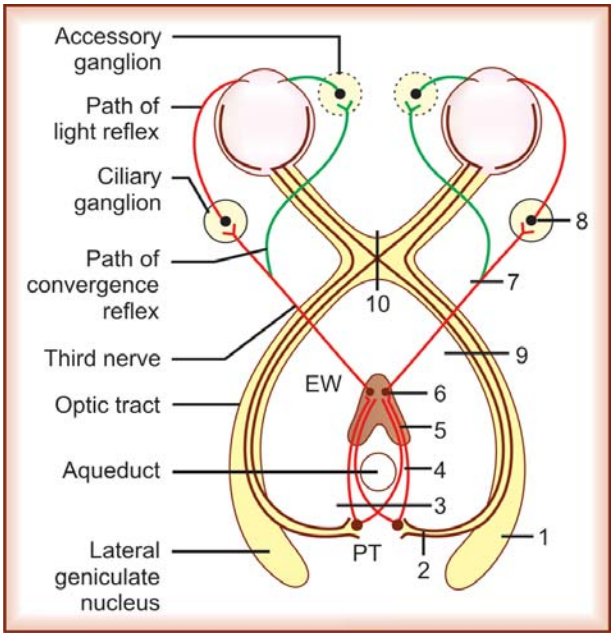

Fig. 3.3: Pathway for light and convergence reflexes (PT—Pretectal nucleus, EW: Edinger-Westphal nucleus)

1. Lesion of proximal part of optic tract: *Normal pupillary reaction*.
2. Lesion in the region of tectum: *Contralateral hemianopic paralysis*.
3. Lesion of central decussation: *Bilateral reflex paralysis*—inactivity to light (direct and consensual) with retention of near reflex, lid reflexes and psychosensory reflex—*bilateral Argyll Robertson pupil* (according to Behr).
4. Lesion between decussation and constrictor center: Ipsilateral abolition of direct and consensual reactions with retention of both contralaterally (*Unilateral Argyll Robertson pupil*).
5. A partial lesion corresponding to 4: Ipsilateral abolition of direct reaction with retention of consensual reaction; retention of both contralaterally.
6. Nuclear or extensive supranuclear lesion: Ipsilateral absolute pupillary paralysis.
7. Lesion of III cranial nerve: Absolute pupillary paralysis.
8. Lesion of ciliary ganglion: Abolition of light reflex with retention of near reflex.
9. Lesion of distal part of optic tract: Contralateral hemianopic paralysis (*Wernicke's hemianopic pupillary reaction*)
10. Lesion of medial chiasma: *Bitemporal hemianopic paralysis.*

contract on convergence. The afferent fibers from these muscles run centrally perhaps through the oculomotor nerve to the mesencephalic nucleus of the trigeminal nerve, to a presumptive convergence center situated in the tectal or pretectal region. Then they reach the Edinger-Westphal nucleus. The efferent impulses run through the III cranial nerve via an accessory ganglion and reach the sphincter pupillae.

Accommodation Reflex

Accommodation reinforces the near reflex along with convergence. The afferent pathway of accommodation is through the optic nerve. The impulse for the accommodation reflex goes with the visual fibers to the lateral geniculate body and then to the striate area of the calcarine cortex to relay in the parastriate area. The efferent fibers travel to nucleus of Perlia via occipito-mesencephalic tract. From Perlia's nucleus fibers go to the Edinger-Westphal nucleus. Then they are carried to the sphincter pupillae muscle through the III cranial nerve via an accessory ganglion.

Psychosensory Reflex

The psychosensory reflex is more complicated and initiated by the stimulation of sensory nerve during pain or emotional states. Sensory excitation initially causes a rapid dilatation of pupil owing to augmentation of the dilator tone via the cervical sympathetics. Then it is followed by a quick second dilatation which lasts longer due to inhibition of the constrictor tone.

The dilator pupillae is supplied by the adrenergic fibers of the cervical sympathetic nerve. Perhaps the tract commences in the hypothalamus and descends downwards through the medulla oblongata into the lateral columns of the spinal cord. The preganglionic fibers leave through the ventral roots of C_8, T_1, T_2 and T_3 nerves and enter the corresponding cord to reach the superior cervical ganglion. From here the postganglionic fibers pass along with the carotid plexus into the skull. Thence, the fibers run along the ophthalmic division of the V cranial nerve, follow the nasociliary nerve and finally reach the dilator pupillae muscle via the long ciliary nerves.

EXAMINATION OF PUPIL

The examination of pupil and pupillary reflexes are described in the chapter on *Examination of the Eye*.

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CHAPTER

4 Elementary Optics

LIGHT

White light (visible sunlight) constitutes a small portion of the electromagnetic spectrum. The visible spectrum ranges from 700 to 400 nm, i.e. from red rays to violet, which the normal eye perceives because of its color sense. The media of the eye are permeable to the visible rays between 600 and 390 nm. The cornea absorbs rays shorter than 295 nm and the lens shorter than 350 nm. The vitreous humor absorbs the rays of about 270 nm. Rays between 400 and 350 nm can reach the retina in normal eye, while those between 400 and 295 nm reach the retina in aphakic eyes. The pigment epithelium of iris and retina absorbs heat radiation in the infrared part of the spectrum from 1100 to 700 nm.

The common objects around us become visible as the light falling on them gets scattered in all directions, while the polished surfaces and mirrors reflect light strongly in a particular direction. The light rays propagate in straight lines and each ray reflected from an object represents the image of the object from which light is reflected. The speed of light (velocity) depends on the optical density of the medium. If the medium is not opaque, a part of the light is reflected back into the first medium and a part of it is refracted.

REFLECTION OF LIGHT

Light rays falling on a surface are *incident rays* and those reflected by it are *reflected rays*. A line drawn at right angles to the surface is called *normal*. The *laws of reflection* are: (i) the incident ray, the reflected ray and the normal at the point of incidence, all lie in the same plane, and (ii) the angle of incidence is equal to the angle of reflection (Fig. 4.1).

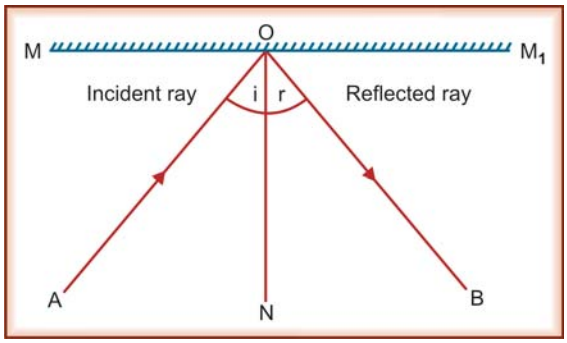

Fig. 4.1: Laws of reflection

Plane Mirror

The image in a plane mirror (Fig. 4.2) is: (i) of the same size as the object, (ii) lies at the same distance behind the mirror as the object is in front, (iii) laterally inverted, and (iv) virtual.

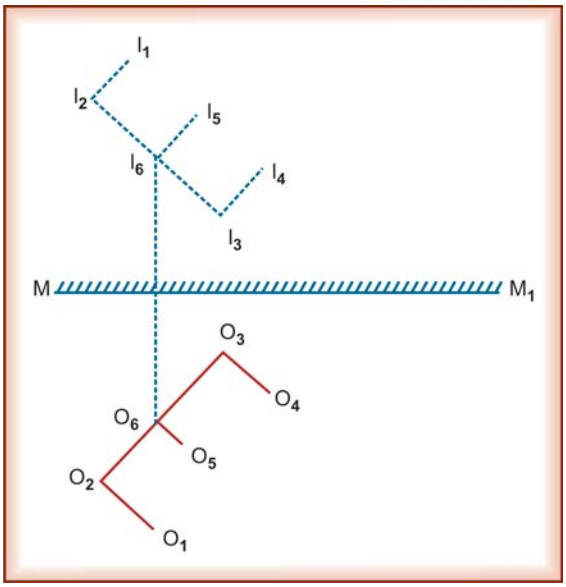

Fig. 4.2: Images in a plane mirror

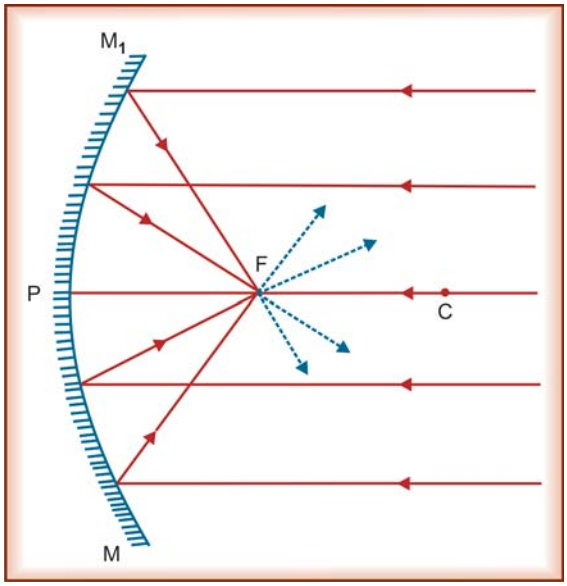

Fig. 4.3: Principal focus of a concave mirror: P—Pole, F—Principal focus, C—Center of curvature, CP—Radius of curvature

Curved Mirror

The *center of curvature* and *radius of curvature* of a spherical mirror are respectively the center and the radius of the sphere of which the mirror was a part. The *pole* of the mirror is the geometric center of the reflecting surface. The *principal axis* of a spherical mirror is the line joining the pole to the center of curvature.

Concave Mirror

The *principal focus* of a concave mirror is that point on the principal axis where light rays traveling parallel to the principal axis converge after reflection. Its *focal length* is the distance between the principal focus and the pole of the mirror and is equal to half the radius of curvature, F = CP/2 (Fig. 4.3).

For a concave or converging mirror, the principal focus is real. The image is real and inverted when the object is between the infinity and the principal focus of the mirror (Fig. 4.4).

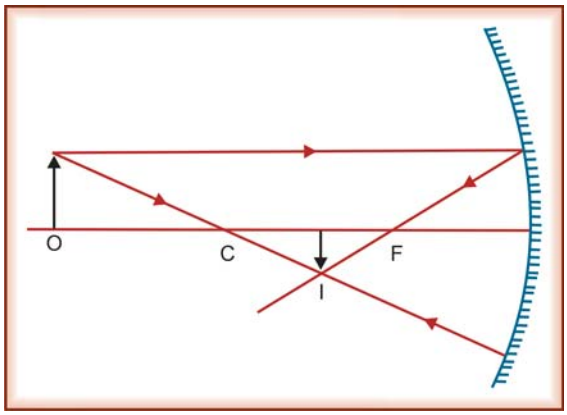

Fig. 4.4: Image formation by a concave mirror: O—Object, I—Image, F—Principal focus, C—Center of curvature

The image is erect, virtual and magnified when the object is between the pole and the principal focus of a concave mirror (Fig. 4.5).

Convex Mirror

The *principal focus* of a convex mirror is that point on the principal axis where the light rays

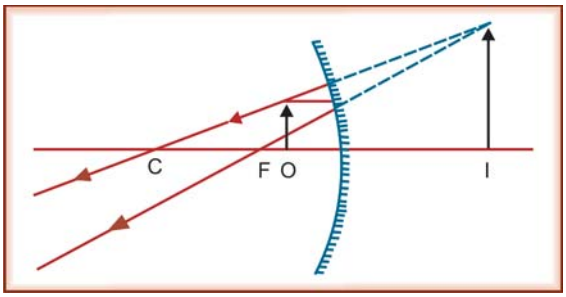

Fig. 4.5: Image formation by a concave mirror: O—object distance less than the principal focus, I—Image erect, virtual and enlarged

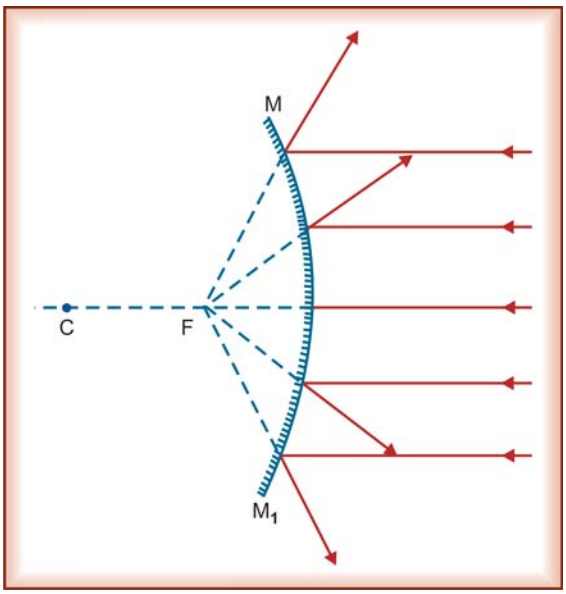

Fig. 4.6: Principal focus of a convex mirror: F—Principal focus, C—Center of curvature, MM_1—Convex mirror

traveling parallel to the principal axis appear to meet after reflection from the mirror (Fig. 4.6).

Unlike the concave mirror which can produce either real or virtual images according to the position of the object, the convex mirror gives virtual images only. These images are always erect and smaller than the size of the object.

The reflection and image formation by curved mirrors is of great importance in ophthalmic optics. The anterior surface of the cornea acts as a convex mirror and this optical property of the cornea is used in keratometry to measure the corneal curvature.

The images formed by the reflecting surface of the eye are called *catoptric images*.

REFRACTION OF LIGHT

When light rays pass through a rectangular slab of glass, the rays are bent or refracted on passing from air to the glass (Fig. 4.7). Simultaneously, a fraction of the light is reflected from the surface of the glass. It is important to remember that when a ray passes from one medium to a more optically dense medium, the ray bends towards the normal. Conversely, a ray passing from a glass or water into air is bent away from the normal.

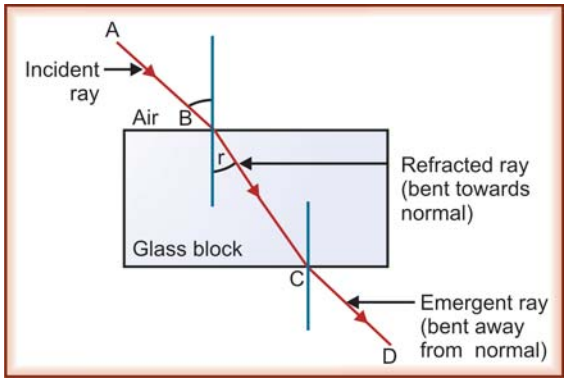

Fig. 4.7: Refraction through parallel sided slab of glass

Laws of Refraction

1. The incident and refracted rays are on opposite sides of the normal at the point of incidence, and all three lie in the same plane.
2. The ratio of sine of the angle of incidence to sine of the angle of refraction is constant. This is also known as *Snell's Law.*

The value of the constant $\frac{\text{sine i}}{\text{sine r}}$ (referred

to in law 2) is called *refractive index* for light passing from the first medium to the second and is denoted by the letter n.

If the first medium is air (or vacuum), then n is called the *refractive index* of the second medium. Refractive index of crown glass is 1.52 (which is used for optical purposes). The refractive index of the flint glass is about 1.65 and that of water is 1.33.

For a ray incident from an optically denser to a rarer medium if the angle of incidence is gradually increased, the angle of refraction also increases but only upto a certain critical value. This angle of incidence is known as *critical angle* (Fig. 4.8) which is the largest angle of incidence for which refraction can still occur.

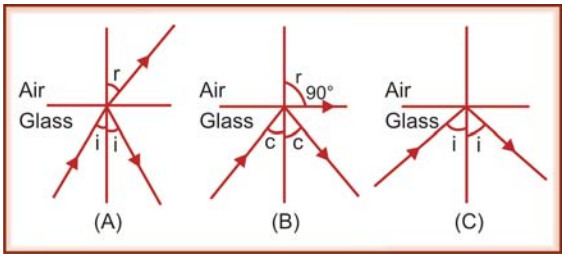

Fig. 4.8: Critical angle (A) refraction and internal reflection i < c, (B) critical refraction i = c, (C) total internal reflection i > c; i: angle of incidence, r: angle of refraction, c : critical angle

In the case of light traveling from one medium to a less optically dense medium, total internal reflection occurs for all angles of incidence greater than the critical angle. The total internal reflection occurs at surfaces within the eye, notably the cornea-air interface.

DISPERSION OF LIGHT

White light is composed of varying wavelengths. The refractive index of any medium differs slightly for light of different wavelengths. Light of shorter wavelength is deviated more than light of longer wavelength.

Prisms

A prism can be described as a portion of a refracting medium (glass or plastic) bordered by two plane surfaces which are inclined at an angle, *apical angle* or *refracting angle of the prism*. Light passing through a prism is refracted according to the Snell's law. The incident ray is deviated towards the base of the prism. The image formed by a prism is erect, virtual and displaced towards the apex of the prism (Fig. 4.9). A prism causes the light to be deviated. The angle of deviation is the angle between the incident and the emergent ray. The deviation is least when the light passes symmetrically through the prism, that is, when the angle of incidence is equal to the angle of emergence. In ophthalmic practice, only thin prisms practise which deviate rays symmetrically are used. The angle of deviation of an ophthalmic prism equals half the refracting angle of the prism. A glass prism of refracting angle 10° (a ten degree prism) deviates the light through 5° and has a power of 10 prism diopters. Prisms are used for the objective measurement of the angle of deviation, measurement of fusional reserve and diagnosis of microtropia, and therapeutically in convergence insufficiency and to relieve diplopia in certain cases of strabismus.

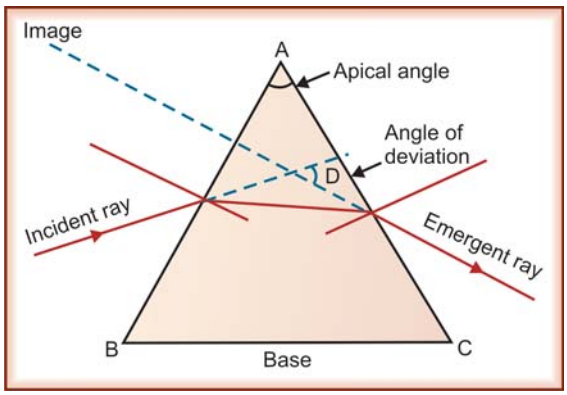

Fig. 4.9: Refraction by a prism

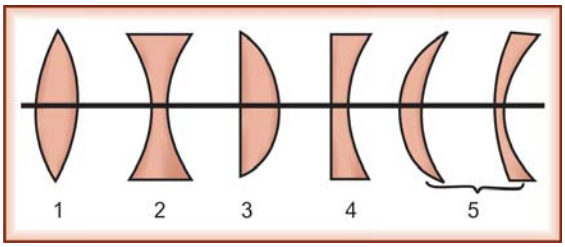

Fig. 4.10: Basic forms of spherical lenses. 1. Biconvex, 2. Biconcave, 3. Planoconvex, 4. Planoconcave, 5. Convex meniscus and concave meniscus

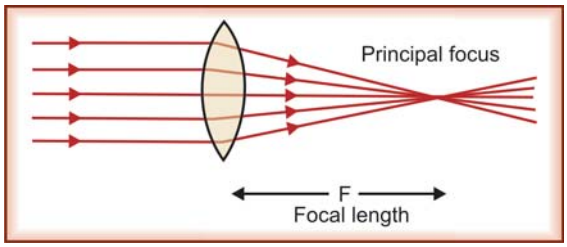

Fig. 4.11: Principal focus of a convex lens

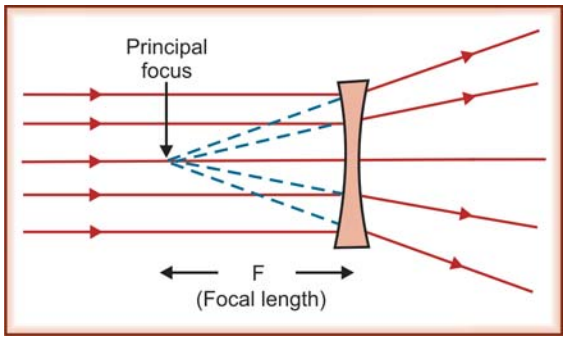

Fig. 4.12: Principal focus of a concave lens

Lenses

A simple lens is usually a piece of glass bounded by spherical surfaces. Figure 4.10 illustrates some of the more common types of lenses. The spherical lenses are divided into two classes.

1. Converging or convex (thickest in the middle), and
2. Diverging or concave (thinnest in the middle).

The *principal axis* of a lens is the line joining the centers of curvature of its faces. The *principal focus* of a lens is that point on the principal axis where all parallel rays of light after passing through the lens converge for a convex lens or they appear to diverge for a concave lens (Figs 4.11 and 4.12). The *focal length* of a lens is the distance between the optical center and the principal focus. The unit of lens power is *diopter* which is reciprocal of the focal length measured in meters. The reciprocal of the second focal length expressed in meters gives the *vergence power of the lens* in diopters (D). A lens which brings the parallel rays to a focus at 1 meter from its optical center is said to have a power of 1 diopter. Lenses of shorter focal length are more powerful than lenses of longer focal length.

A convex lens produces a real inverted image if the object is placed at a distance greater than the focal length of the lens and a virtual erect image when the distance is shorter than the focal length (Figs 4.13 and 4.14).

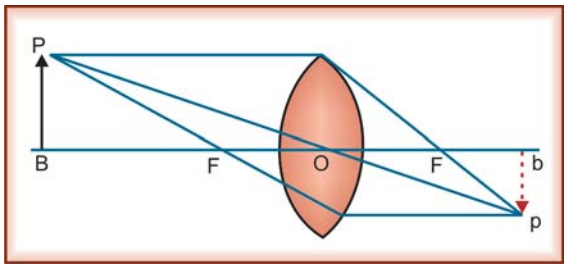

Fig. 4.13: Image formation by a convex lens. bp: real image, PB: object beyond F

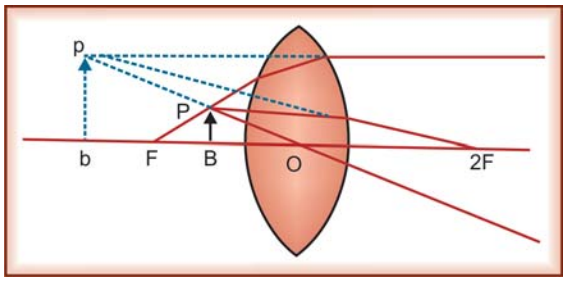

Fig. 4.14: Image formation by a convex lens. bp: virtual image, PB: object between lens and F

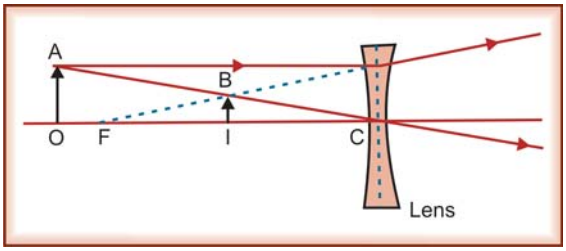

Fig. 4.15: Image seen in a concave lens.
OA: object, IB: virtual image

A concave lens always produces a virtual, erect and smaller image than the object (Fig. 4.15).

A convex lens can be considered as a series of prisms with their bases together and a concave lens with their bases away. Therefore, light rays are deviated more at the edges of a lens than at the center. This explains the convergence and divergence of the rays, respectively.

When two lenses of opposite sign but equal curvature are placed in contact with one another, the resultant effect will be that of a plate with parallel sides which does not deviate the rays of light. Thus, the power of an unknown lens can be found by neutralizing it with a lens of the opposite sign.

System of Lenses

When more than one lens is used, the refractive power of the combination will be equal to the algebraic sum of the powers of these lenses provided that the lenses are thin and centered on a common optical axis.

If the lenses in an optical system are thick or placed at a distance separated by a medium, the resultant power of the system will evidently differ from the effect of thin lenses placed together. The resultant focal power of such a system of lenses can be found by the expression:

$$F_v = (F_1 + F_2)\ t/n\ (F_1)^2$$

where, F_1 and F_2 are the front and back surface powers or the power of the first lens and the second lens, respectively, t is the thickness or the distance separating the lenses and n is the refractive index of the lenses.

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CHAPTER

5 Physiological Optics

The cornea, the aqueous humour, the lens and the vitreous constitute the optical system of the eye and bring the rays of light to a focus upon the retina.

Reduced Schematic Eye

The optical system of the eye is quite complicated. To conceptualize and understand the optical properties of the human eye, it can be simplified theoretically as one convex lens (cornea) having a power of 60 diopter separating the two media of refractive indices of 1 and 1.33. This lens is placed at a principal point P with one optical center, the nodal point N, which lies about 5.6 mm behind the anterior surface of the cornea. The anterior focal point (F_1) is situated nearly 17 mm in front of the cornea and the posterior (F_2) approximately 22.6 mm behind the cornea coinciding with the position of the retina in a normal eye (Fig. 5.1). Such an optical system is called a *reduced schematic eye*. It allows to determine the sizes of retinal lesions, calculation of intraocular lens power for implantation and localization of intraocular foreign body.

In some eyes, the retina is not situated at its usual position and, therefore, the parallel rays from a distant object may be focused either in front or behind the retina. The former condition is called *myopia* and the latter *hypermetropia (hyperopia)*.

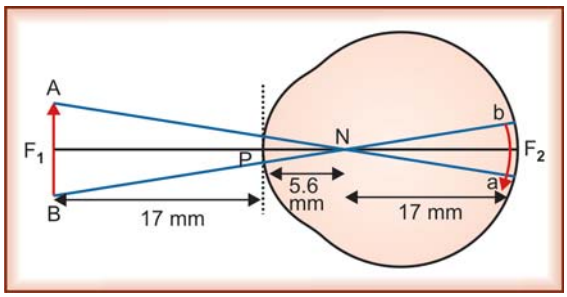


Fig. 5.1: The reduced schematic eye

Both the conditions are collectively called *ametropia* or *errors of refraction*; in each condition a blurred image is formed upon the retina and the vision remains subnormal. If the refraction of the two eyes are different, the condition is called *anisometropia*.

Consider the light rays from the object AB passing through the nodal point and reaching the retina to form the image ab (Fig. 5.1). The size of the retinal image depends upon the angle subtended by the rays at the nodal point and also on the distance of the retina from the nodal point. If the retina is located nearer to the nodal point, as seen in hypermetropia, the size of the blurred image is smaller than that formed in emmetropia. On the other hand, the nodal point lies farther from the retina in the myopic eye, hence, the image size is larger.

The hypermetropic eye is usually small and the rays coming from a point on the retina appear more divergent than the corresponding rays from emmetropic eyes (compare the effect of placing an object closer to a convex lens than its principal focus). They meet behind the eye if produced backward (Fig. 5.2), thus the far point (*punctum remotum*) of the hypermetropic eye is virtual and lies behind the eye. The nearer the retina is to the lens, the higher the degree of hypermetropia. The point on the retina and the far point are conjugate.

Consider the image formation in a myopic eye which is too long. The rays emerging from a point on the retina are less divergent than the corresponding rays in emmetropic eyes (compare the effect of placing an object farther away from a convex lens than its principal focus). They cross at a point somewhere in front of the eye (Fig. 5.3). Therefore, the far point of the myopic eye is real and lies in front of the eye. The far point and the point upon the retina from where the rays emerge are conjugate to each other. The farther the retina is to the lens, the higher the degree of myopia and nearer lies the far point as the emergent rays are more convergent.

It is evident that in every case the far point and a point on the retina are in conjugate focus. Considering the reversibility of the rays, the object situated at the far point of the eye has a sharp image upon the retina. Emmetropic eye can see the object clearly up to infinity (objects more than 6 meters away). Myopes can see the object clearly

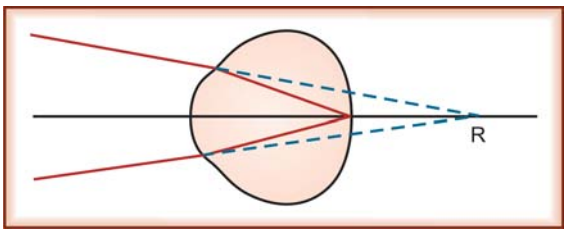


Fig. 5.2: Hypermetropic eye: the emergent rays diverge apparently from a point R behind the eye

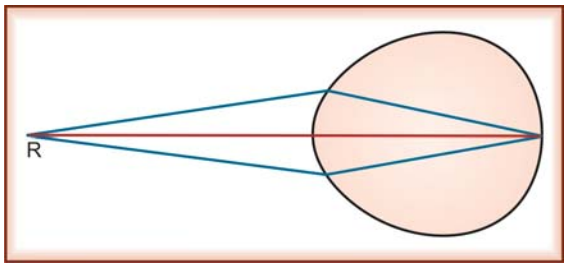


Fig. 5.3: Myopic eye: the emergent rays converge at a point R in front of the eye

located only near to the eye, hence, termed as *short-sighted*. The hypermetrope cannot see either the distant or the near object clearly unless the individual makes an effort of accommodation.

Errors of refraction or ametropia may be due to several factors.

1. Axial length of the eye
2. Refractive index of the media, and
3. Curvature anomalies.

Axial ametropia results from undue shortening or lengthening of the eye. *Index ametropia* occurs due to alteration in the refractive indices of the media and *curvature ametropia* is caused by alteration in the curvature of the cornea or the lens. If the curvatures of the two principal meridians (horizontal and vertical) are different, the condition causes a troublesome error of refraction, known as *astigmatism*. The astigmatism may be of two types—*regular* and *irregular*. When the cornea has its direction of the greatest and the least curvature at right angles to one another it is termed as *regular astigmatism*. Owing to the pressure from the upper lid the vertical meridian is more curved, it is called *regular astigmatism with-the-rule*. The reverse condition is known as *regular astigmatism against-the-rule*. The irregularity of the corneal surface causes distortion of the meridians resulting in irregular refraction of light rays that get focused at various positions, such a condition is known as *irregular astigmatism*.

Sturm's conoid is an image produced by an astigmatic eye and represents the pattern of rays

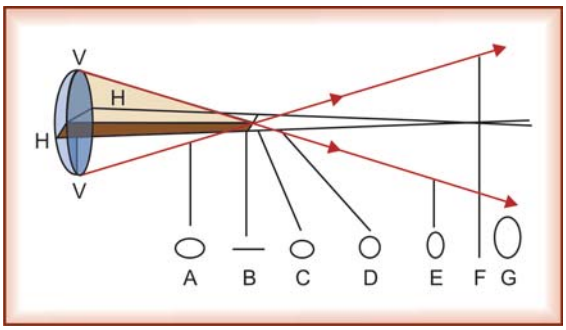


Fig. 5.4: Image formation by an astigmatic eye (Sturm's conoid). VV: corneal vertical plane, HH: corneal horizontal plane. ABCDEFG illustrate nature of the image in different positions of retina in relation to cornea.

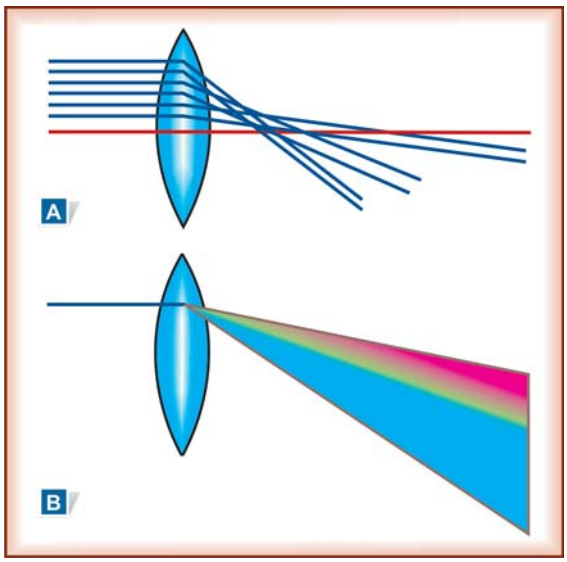
formed after passage through a spherocylindrical combination. According to the position of the retina in relation to the focal lines, astigmatism can further be divided into following types (Fig. 5.4).

1. If the retina is positioned at A, both the focal lines form focus behind it. This condition is called *compound hypermetropic astigmatism*.
2. In position B, the horizontal focal line formed by the vertical meridian is on the retina and the other focal line is behind it. This condition is known as *simple hypermetropic astigmatism*.
3. In position (C, D and E) one focal line is in front and another behind the retina giving rise to *mixed astigmatism*. In position D, the converging and the diverging light rays meet forming a circular and clear image known as *circle of least diffusion*.
4. In position F, the vertical focal line formed by the flatter horizontal meridian is in focus on the retina. This condition is called *simple myopic astigmatism*.
5. In position G, both the focal lines form a focus in front of the retina, this condition is called *compound myopic astigmatism*.

ABERRATION IN RETINAL IMAGES

The optical system of the eye is by no means perfect. The imperfections in the nature of retinal images are described as *aberrations*. They are of two types—spherical and chromatic. The *spherical aberration* occurs due to difference in the central and the peripheral curvature of the cornea. The light rays nearer to the principal axis (paraxial) are brought to a sharp focus, while the peripheral rays form overlapping images causing blurring (Fig. 5.5A). The iris, however, acting as a diaphragm cuts off the peripheral rays and minimizes the defect.

White light is composed of all the colors of the spectrum. The light rays of longer wavelength (red end) are refracted least and of shorter wavelength (violet) most, causing *chromatic aberration* (Fig. 5.5B).



Figs 5.5A and B: Aberrations: (A) Spherical, (B) Chromatic

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CHAPTER

6 Errors of Refraction

When parallel rays of light from infinity come to a focus on the retina with accommodation at rest the condition is called *emmetropia* (Fig. 6.1). Conversely, when the parallel rays of light from infinity do not come to a focus upon the retina with accommodation at rest it is known as *ametropia*.

Ametropia may be due to following causes.

1. *Abnormal length of the globe—axial ametropia* wherein too long and too short lengths of the globe result in myopia and hypermetropia, respectively. Perhaps, the change in the axial length of the globe is the most important cause of ametropia.
2. *Abnormal curvature of the cornea or the lens—curvature ametropia* wherein too much and too less curvatures cause myopia and hypermetropia, respectively.
3. *Abnormal refractive indices of the media—index ametropia* wherein increase in the indices of the refractive media (cornea, aqueous and lens) and decrease in the index of vitreous cause myopia, while the opposite conditions lead to hypermetropia.
4. *Abnormal position of the lens*—a forward displacement of the lens leads to myopia and backward displacement to hypermetropia.

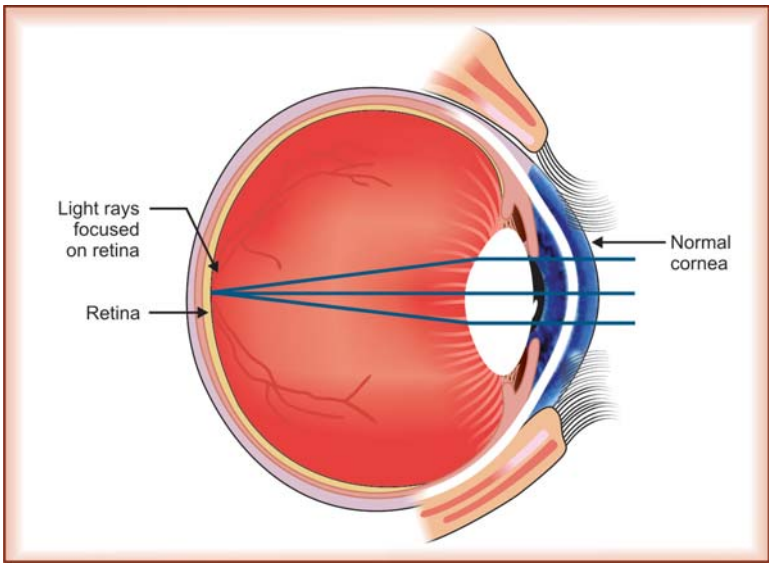

Fig. 6.1: Emmetropia

There are three types of errors of refraction: (i) *myopia* or short-sightedness, (ii) *hypermetropia* or long-sightedness, and (iii) astigmatism.

MYOPIA

Myopia is that diopteric condition of the eye in which parallel rays of light from infinity come to a focus in front of the retina when accommodation is at rest (Fig. 6.2).

At birth most eyes are small and hypermetropic, but as the growth proceeds they increase in size (to reach the normal adult size of 24 mm) and become emmetropic. If the lengthening of the eyeball continues, *axial myopia* results (1 mm = 3 D). *Curvature myopia* commonly occurs due to an abnormal curvature of the cornea as seen in keratoconus, and less frequently because of increased curvature of the lens, lenticonus (0.1 mm = 3D). *Index myopia* is found in nuclear sclerosis not uncommon in old age.

Types of Myopia

Clinically, myopia is classified as below.

1. Development myopia
2. Simple myopia, and
3. Pathological myopia.

Developmental Myopia

It is rare and characterized by an abnormally long eyeball at birth having a refractive error of 10 D. The fundus shows marked choroidal sclerosis, hypopigmentation and myopic crescent. The developmental myopia is stationary and progression is quite rare.

Simple Myopia

It is the commonest type of myopia which progresses during childhood and adolescence and seldom exceeds 5 to 6 D. It generally stops to progress by the age of 21 years and the best

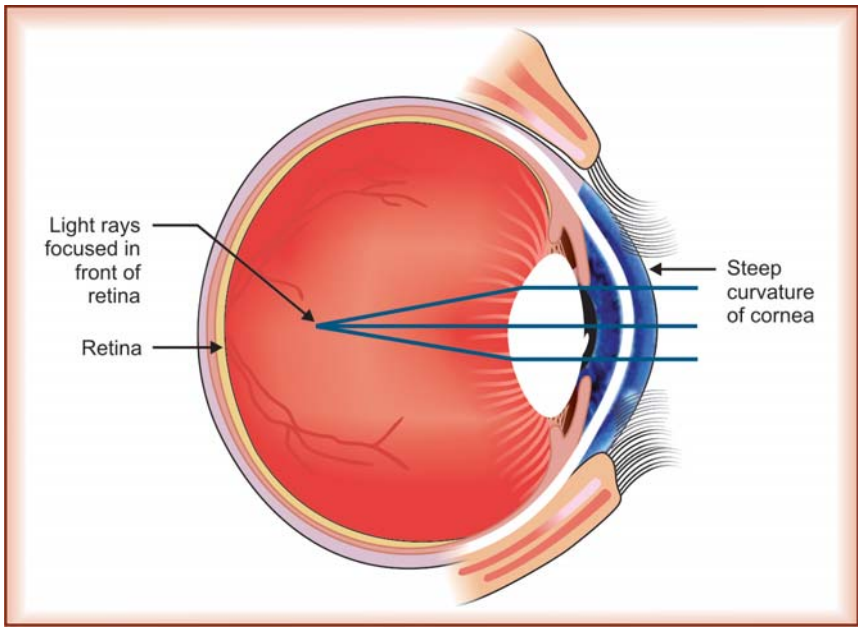

Fig. 6.2: Myopia

corrected visual acuity is always normal (6/6). The fundus may show myopic crescent at the temporal margin of the disk, tigroid fundus and lattice degeneration with or without a retinal break.

Transient and *acquired myopia* may be found following trauma to ocular structures, intraocular lens implantation (over-correction of aphakia), administration of certain drugs (acetazolamide, oral contraceptives, tetracycline, sulfonamides, etc.) and spasm of accommodation (*pseudomyopia*).

Pathological Myopia

Pathological myopia is essentially a degenerative and progressive condition which manifests in early childhood. The refractive error rapidly increases during the period of active growth and may reach 20 to 30 D by the age of 25 years. The condition has a strong hereditary tendency and is more common in women than in men. Autosomal dominant pathological myopia has been linked to genes 18p11.31 and 12q2123. The elongation of eyeball occurs primarily due to degeneration of the posterior half of sclera and is often accompanied with an outward scleral bulge at the posterior pole—*posterior staphyloma*.

A myopic eye has its *punctum remotum* between infinity and the eye and it accommodates less than emmetropic and hypermetropic eyes.

Clinical Features

The inability to see distant objects clearly and holding the book too close to the eye while reading are the usual complaints of parents of the child having simple myopia. Eyestrain and headache may occur due to an imbalance between accommodation and convergence in myopia. Sometimes, the patient sees black spots floating before the eyes and occasionally flashes of light are noticed.

In pathological myopia, the eyes are unusually prominent with slightly dilated pupils. There may be an apparent convergent squint due to a large negative angle kappa. In spite of the optical correction, the vision is poor. The blind spot is enlarged and peripheral visual field is generally constricted.

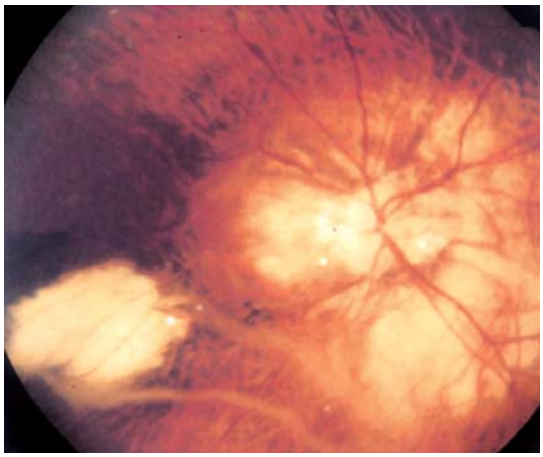

Fig. 6.3: Myopic crescent with chorioretinal degeneration

Ophthalmoscopy may reveal vitreous degeneration and opacities, a big optic disk with myopic crescent and nasal supertraction due to extension of retina on the nasal side of the disk. The crescent may run all around the disk in an annular ring (Fig. 6.3). Besides, there are chorioretinal atrophic patches at the posterior pole as well as in the periphery of the fundus. Choroidal sclerosis and Foster-Fuchs spot at macula due to choroidal hemorrhage may be found. A highly myopic eye is prone to develop retinal hemorrhages, due to complicated posterior vitreous detachment, and lattice degeneration with retinal holes and/or tears leading to detachment of retina and complicated cataract.

Treatment

The treatment of myopia comprises prescribing appropriate concave lenses (Fig. 6.4) and paying attention to ocular hygiene. Generally, the myopia must never be over-corrected and in practice high myopia is almost always slightly under-corrected.

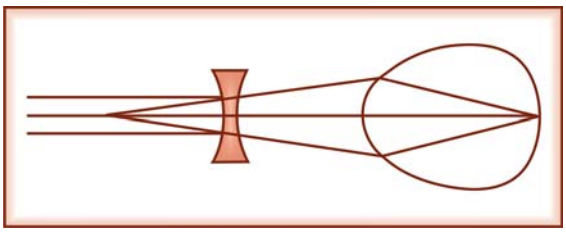

Fig. 6.4: Myopic eye. Parallel rays are brought to a focus on the retina by an appropriate concave lens

Simple myopia up to 6 D may be fully corrected and the patient is advised to do near work at ordinary reading distance. If any discomfort is experienced, weaker glasses may be ordered for near work.

The children with uncorrected myopia may lose interest in their surroundings owing to blurred vision. Hence, the glasses must be worn constantly. In high myopia, the patient often sees best with under-correction as strong concave lenses considerably diminish the size of retinal image. Sometimes, very bright and clear images are not tolerated by the patient whose retina has become accustomed to large and blurred images.

Contact lenses are very helpful in many cases of high myopia. They also eliminate the peripheral distortion caused by thick concave lenses. At the same time, a minus-edge lenticular design of contact lens decreases the discomfort caused by the thickened skirt.

High axial myopia of about 21 D may be corrected by the removal of the crystalline lens, though it is not free from complications owing to the fluidity of vitreous and retinal degeneration. Recently, refractive surgeries have been advocated for the correction of myopia. They include radial keratotomy (RK), photorefractive keratectomy (PRK), laser-assisted epithelial keratomileusis (LASEK) and intracorneal rings (ICR) for mild to moderate degree of myopia (1 to 6 D) and laser-assisted *in situ* keratomileusis (LASIK) for correction of myopia between 2 and 16 D.

In high myopia, the normal relationship between accommodation and convergence is disturbed and if the glasses are not constantly used, the effort to converge is practically abandoned. Thus, the patient uses only one eye for near work and the other eye becomes divergent due to disuse.

The general health of a myopic child should always be attended to. Nutritious diet, outdoor activities and regular exercises should be encouraged. The individual should be advised to do near work in good illumination and continuous reading, particularly at night hours, be discouraged. Should the patient be ill, all near work is stopped otherwise myopia increases rapidly.

In pathological myopia, glasses or contact lenses seldom improve the vision to normal as degenerative changes affect the retina. Low vision aid may be of some help to the patient, particularly in reading. There is no treatment to stop the increase in axial length of the eyeball and arrest the progression of pathological myopia. As these patients invariably develop retinal and macular complications, routine monitoring for retinal break and choroidal neovascular membrane formation is required. The only viable refractive surgery in these cases is either clear lens extraction or phakic intraocular lens implantation. Genetic counseling may stop hereditary propagation of the disease. High myopes with progressive degeneration of the retina should be asked to avoid contact sports or activities as they increase the risk of retinal detachment.

HYPERMETROPIA (*HYPEROPIA*)

Hypermetropia is an error of refraction wherein parallel rays of light from infinity come to a focus behind the retina when accommodation is at rest (Fig. 6.5). Like myopia, the hypermetropia may be *axial, curvature* and *index*. When the antero-posterior length of the globe is shorter than the

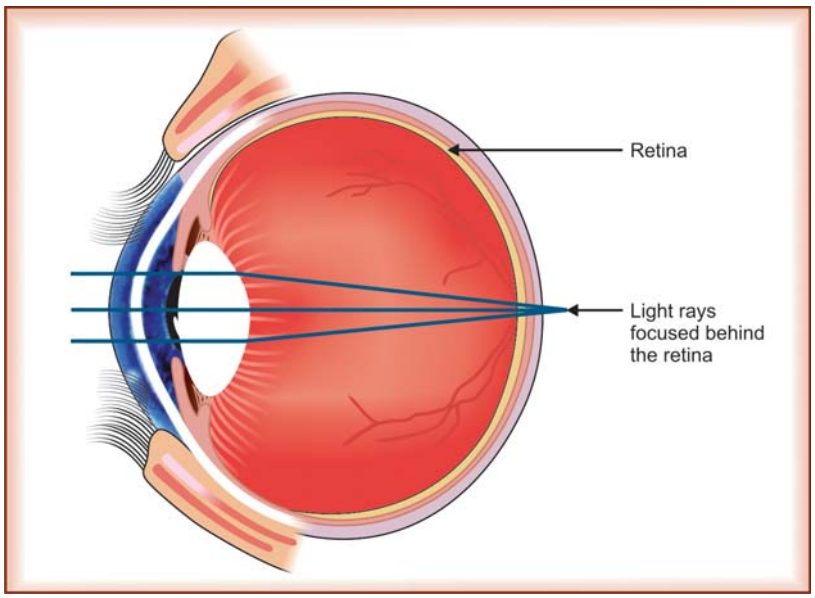

Fig. 6.5: Hypermetropia

normal, *axial hypermetropia* results. Hypermetropic eye is usually smaller in all dimensions than the normal eye; 1 mm shortening of the eye leads to 3 D of refractive error. Almost all eyes at birth are hypermetropic and with the growth of the body their anteroposterior diameter increases and reaches normal length in adolescence. If an eye remains under-developed, hypermetropia is often found. If the curvature of the cornea or lens is flatter than normal, *curvature hypermetropia* occurs. Astigmatism is usually accompanied with curvature hypermetropia. The total decrease in the refractive index of the lens, as found in cortical cataract, accounts for *index hypermetropia*. A backward dislocation of the lens produces hypermetropia. Aphakia (absence of the lens) is an example of a high degree of hypermetropia.

Types of Hypermetropia

Accommodation has a considerable influence on hypermetropia. Depending upon the act of accommodation *total hypermetropia* may be divided into following types:

1. *Latent hypermetropia* which is corrected by the physiological tone of the ciliary muscle.
2. *Manifest hypermetropia* is made up of following two components.
   a. *Facultative hypermetropia* is that part of the error which can be corrected by an effort of accommodation, and
   b. *Absolute hypermetropia* which cannot be overcome by either accommodation or ciliary tone.

Clinically, the types of hypermetropia can be assessed. Generally, a hypermetrope cannot see a distant object clearly unless he accommodates. If convex lenses of gradually increasing strength are placed in front of the patient's eyes until he just sees the object clearly with the weakest convex lens (convex lens and accommodation both acting to provide a clear vision), the amount of hypermetropia corrected by the lens (not corrected by the effort of accommodation) is the *absolute*

hypermetropia. Now place convex lenses of gradually increasing strength until the clear vision is still maintained with the strongest convex lens. This process measures the amount of hypermetropia which the patient corrects by his accommodation, the *facultative hypermetropia*. It is determined by the difference between the strongest and the weakest convex lens, while the strongest convex lens is the measure of *manifest hypermetropia*. Topical cycloplegic is used to paralyse the ciliary muscle. The reafter, the strongest convex lens is placed with which maximum visual acuity can be obtained. It represents the *total hypermetropia*. The amount of *latent hypermetropia* can be worked out by subtracting the manifest hypermetropia from the total hypermetropia.

Clinical Features

Low degree of hypermetropia may not cause any symptoms in young individuals as they have ample reserve of accommodation. However, symptoms may appear with the decline of accommodation in later life. In high hypermetropia, the available accommodation may not adequately cope with the error, hence, blurring of vision may occur for distance as well as for near. Symptoms are often aggravated by long continued close work or reading. Headache is a common sequel to the excessive accommodation needed for near work. The overaction of ciliary muscle is likely to produce eyestrain. The condition is known as *accommodative asthenopia*. Heaviness of the lids, dull pain in the eye and congestion of the eye are the other symptoms. Young hypermetropes are prone to develop latent convergent squint which further increases the eyestrain. In general, presbyopia commences at an early age than usual in hypermetropes. A hypermetropic eye is usually smaller than the normal, particularly along the axial length. The diameter of the cornea is reduced and the anterior chamber is often more shallow than usual. Such an eye is predisposed to angle-closure glaucoma. A bright reflex resembling a watered-silk or shot-silk appearance may be found in hypermetropia on funduscopy. Occasionally, the margin of the disk may be seen blurred, *pseudopapillitis*, and the blood vessels may be unduly tortuous.

Treatment

Hypermetropia with asthenopia is corrected by prescribing convex lenses (Fig. 6.6). In young children with heterophoria, examination should be conducted under a cycloplegic. One diopter is additionally deducted from the retinoscopy to allow for the ciliary tone and the prescribed glasses must be used constantly. In these children, hypermetropia tends to diminish with growing age, hence, they must be examined once a year for a possible change in their glasses. In young patients with active accommodation hypermetropia should be undercorrected but in advanced age, when all the manifest hypermetropia becomes absolute and accommodation is poor, a full correction is advised. Mild to moderate degree of hypermetropia (1 to 4 D) can be managed by LASIK or conductive keratoplasty while high degree of hypermetropia can be corrected by phakic intraocular lens implantation. The cases of aphakia are increasingly being managed by secondary intraocular lens implantation.

ASTIGMATISM

Astigmatism is that condition wherein the refraction varies in different meridians of the eye. Hence, a point focus cannot be formed upon the retina (Fig. 6.7). Astigmatism is most commonly caused by abnormalities in the curvature of the cornea (*curvature astigmatism).* Abnormalities in the curvature or centering of the lens can also cause astigmatism. A small amount of astigmatism occurs due to inequalities in the refractive index of different sectors of the lens (*index astigmatism*).

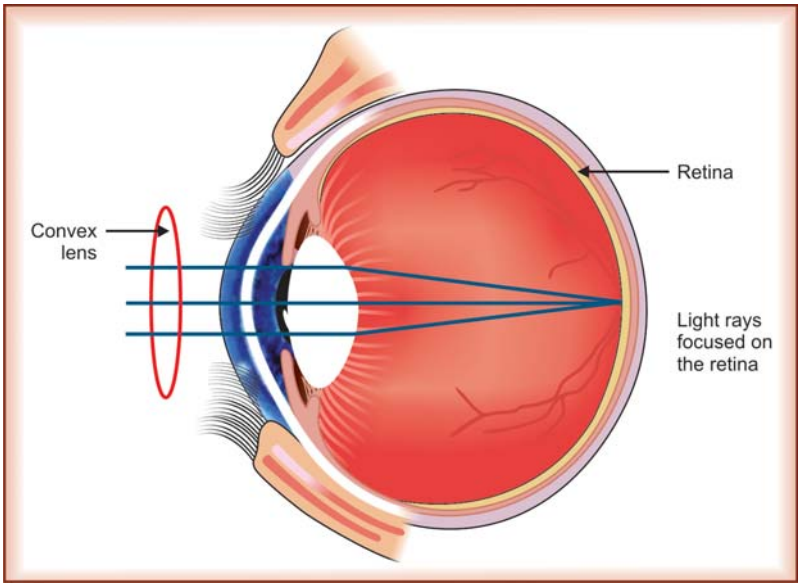

Fig. 6.6: Hypermetropic eye. Parallel rays are brought to a focus on the retina by an appropriate convex lens

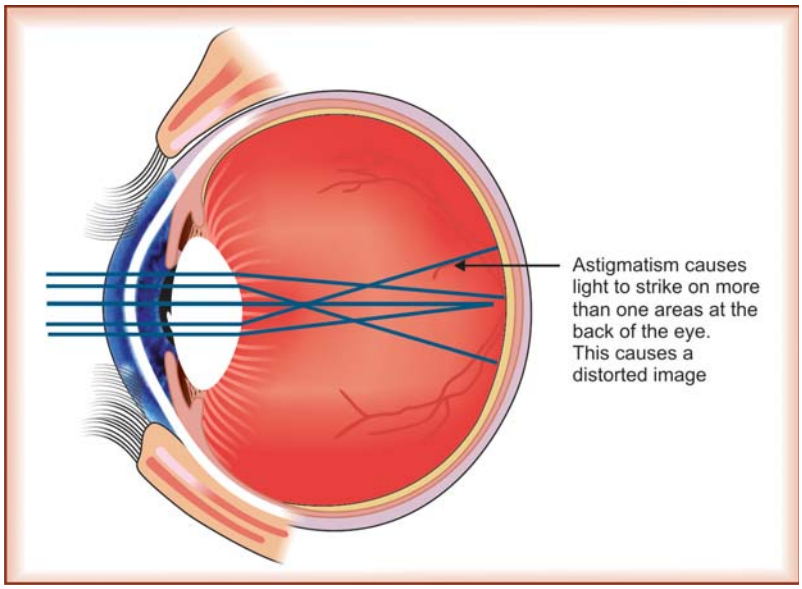

Fig. 6.7: Astigmatism

Types of Astigmatism

Theoretically, no eye is stigmatic as the vertical curvature of the cornea is greater than the horizontal by about 0.25 D owing to the pressure of the upper lid upon the eye. This is accepted as physiological and termed as *astigmatism with-the-rule*. As age advances, it tends to disappear or even gets reversed to *astigmatism against-the-rule*

wherein the horizontal curvature becomes greater than the vertical. The most common cause of astigmatism against-the-rule is cataract surgery from superior corneal, limbal or corneoscleral section in which the vertical meridian flattens due to the scarring.

Broadly speaking, astigmatism is divided into two categories—*regular* and *irregular*. When the two principal meridians of greatest and least curvature are at right angles to each other, the condition is called *regular astigmatism*. Occasionally, the axes are not at right angles but are crossed obliquely; this condition is known as *bi-oblique astigmatism*. If the two meridians do not lie in the principal planes (that is near to 90 or 180 degrees), but remain at right angles to each other, this type of regular astigmatism is termed as *oblique astigmatism*.

Regular Astigmatism

Regular astigmatism may be classified into the following types:

1. *Simple astigmatism*, where one of the principal meridians is emmetropic and the other is either hypermetropic or myopic. The former is known as *simple hypermetropic* and the latter *simple myopic astigmatism*.
2. *Compound astigmatism*, where both the principal meridians are either hypermetropic or myopic, the former is known as *compound hypermetropic* and the latter *compound myopic astigmatism*.
3. *Mixed astigmatism*, where one of the principal meridians is hypermetropic and the other myopic.

Clinical Features

Generally, small astigmatic errors do not give any ocular discomfort. However, severe symptoms are found in cases of hypermetropic astigmatism wherein the accommodation is brought into play to overcome hypermetropia. Higher degrees of astigmatism often cause poor visual acuity but vision is not much impaired in mixed astigmatism as the circle of least diffusion falls upon or near the retina. The continuous strain of accommodation may cause symptoms of asthenopia. The optic disk appears oval or blurred in one sector in astigmatism on direct ophthalmoscopy.

An *astigmatic fan*, consisting of horizontal and vertical lines may help to detect the regular astigmatism. The patient sees distinct lines of the fan in one direction (vertical or horizontal) and they appear tailed off or blurred in the other direction.

Irregular Astigmatism

When the curvature and refractive power of the refractive media are markedly irregular causing multiple focal points which produce completely blurred images on the retina such a condition is called *irregular astigmatism.*

The irregular astigmatism is caused by corneal scar, penetrating injuries of the eye, keratoconus, lenticonus and immature cataract. The patient with irregular astigmatism often suffers from distorted vision and headache.

Treatment

A small degree of astigmatic error may not require any optical correction. But in all such cases, if the error causes asthenopic symptoms, a full optical correction by cylindrical lenses should be advised for constant use. All forms of regular astigmatism can be corrected by cylindrical lenses or spherocylindrical combinations. In contrast, irregular astigmatism cannot be corrected by spectacle lenses due to irregularities in curvature of meridians.

The visual acuity does not improve in cases of irregular astigmatism with spectacle correction; here toric contact lenses are of immense value.

Soft or rigid gas permeable toric lenses are prescribed.

Large degrees of astigmatism following cataract extraction and keratoplasty can be managed by laser *in situ* keratomileusis or conductive keratoplasty. Astigmatic keratotomy, relaxing incisions in the cornea, or limbal relaxing incision can correct mild astigmatism. In all these techniques the closer the incision to the center of the cornea, the greater its influence on astigmatism.

ANISOMETROPIA

Anisometropia is that condition wherein there is relative difference in the refractive status of the two eyes. It is significant when the difference between the refraction of the two eyes exceeds 2.5 D. A minor difference of refraction between the two eyes is not uncommon and it seldom gives any symptom. Binocular vision is usually maintained if the difference between the two eyes does not exceed 2.5 D. In some cases, when one eye is emmetropic or moderately hypermetropic and the other is myopic, the patient falls into the habit of using emmetropic or hypermetropic eye for distant vision and the myopic for near work. The binocularity is disrupted in high degrees of error as the patient tends to suppress the image in the more ametropic eye. It ultimately leads to *amblyopia ex-anopsia* (amblyopia due to disuse). High degree of uniocular myopia, hypermetropia and uniocular aphakia are important causes of anisometropia.

Treatment

Anisometropia must be corrected in childhood to prevent the development of amblyopia ex-anopsia. The optical correction is not readily acceptable to the child due to difference in the size of the retinal images (*aniseikonia*) in the emmetropic and the corrected eye. The use of contact lens eliminates this defect. If the eye has become amblyopic, the emmetropic eye is patched and the patient is encouraged to use the ametropic eye with optical correction. Later, orthoptic exercises should be given to develop binocularity. Laser *in situ* keratomileusis has been tried with satisfactory results in cases of anisometropia.

ASTHENOPIA

Asthenopia is characterized by ocular or periocular discomfort, heaviness of eyelids, sleepiness, tired eyes, browache and headache associated with prolonged ocular use especially for near. Occasionally, the patient complains of throbbing headache often accompanied with nausea.

The main causes of asthenopia are:

1. Uncorrected refractive errors—hypermetropia and astigmatism
2. Incorrect glasses or misplacement of the optical center of a corrective lens
3. Heterophorias
4. Anisometropia
5. Presbyopia
6. Convergence deficiency.

Treatment

The causes of asthenopia must be identified and should be treated promptly. The corrective measures include correction of refractive error, replacement of inappropriate glasses, orthoptic exercises and/or surgical correction of muscle imbalance.

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CHAPTER

7 Determination of the Refraction

The refractive errors are determined in practice, both objectively and subjectively. The objective determination of the refraction can be done either by *objective optometry* or *retinoscopy* and occasionally by *keratometry*.

AUTOREFRACTOMETERS

Autorefractometers (Fig. 7.1) are employed for objective determination of refractive errors. The optometers are automated computerized instruments which measure quickly the far point of the eye and give instantaneous printout of the refractive error of the subject screened in terms of sphere, cylinder, axis, interpupillary distance and other technical data. However, these readings cannot be blindly prescribed, as the subjective acceptance and tolerance significantly differ in practice owing to personal and instrumental errors. Autorefractometers can be considered logically as substitutes for retinoscopy. They can be used advantageously for mass screening, research programs and epidemiological studies.

Fig. 7.1: Nidek autorefractometer
(Courtesy: Biomedix, Bangalore)

RETINOSCOPY

The most commonly employed method for determining the refraction objectively is *retinoscopy* or *skiascopy*. The basic principle of retinoscopy is that when light is reflected from a mirror into an eye, the direction in which the light travels across the pupil varies with the refraction of the eye. The retinoscopy is a process by which the far point of the eye is brought nearer to the eye, say at 1 meter, with the help of trial lenses placed in front of the eye under examination.

It has already been explained that the emergent light rays are parallel in emmetropic, divergent in hypermetropic, and convergent in myopic eyes. We know that the far point of the myopic eye (1D) lies at 1 meter in front of the eye and the trial lenses can be employed to alter the vergence in front of the eye.

Basically, the instruments needed for objective retinoscopy are a light source and a mirror, plano or concave (Fig. 7.2), or a self-illuminated spot or streak retinoscope (Fig. 7.3). The patient is seated at about 1 meter distance. His fundus is illuminated with the help of a mirror held by the examiner in his hand. The reflected light from the fundus is seen as a red reflex filling the pupillary area. If the mirror is tilted either upwards, downwards or to right or left, the reflex also appears to move. This illusionary movement of the reflex with the movement of the mirror depends upon the nature of the refractive status of the eye.

If a plane mirror is used for retinoscopy at a working distance of 1 meter, the movement of reflex is in the same direction (*with movement*) to the movement of mirror in emmetropia, hypermetropia and myopia of less than 1 D, whereas the reflex moves in the opposite direction to the movement of the mirror in myopia of more than 1 D.

If a concave mirror is used instead of a plane mirror and the other conditions remaining the same, the reflex moves in the opposite direction (*against movement*) to the movement of mirror in emmetropia, hypermetropia and myopia of less than 1 D but reflex moves in the same direction to the movement of the mirror in myopia of more than 1 D.

If an eye has a myopia of 1 D, the retinoscopic reflex at a distance of 1 meter with a plane mirror becomes neutral, i.e. there is neither with movement nor against. Either the reflex appears bright or completely dark. The pupillary reflex, with a slight tilt of the mirror, disappears quickly and completely. This point is called *neutral point* or *neutral reflex*. An optometrist, while doing refraction, attempts to obtain this point by adding appropriate lenses in the trial frame. It is indeed difficult to obtain the neutral point, hence, a *point of reversal* is achieved. This condition is created by adding a slightly higher power (+ 0.25 D) than

Fig. 7.2: Retinoscope—plano and concave mirrors

Fig. 7.3: Streak retinoscope

the existing refractive error. Generally, –1 D is added to the power of trial lenses in order to calculate the actual refractive error. For example, in a hypermetropic individual if the point of reversal is achieved with + 4 D sphere, the actual error is (algebraic sum of + 4 D sphere – 1 D sphere for a working distance of 1 meter) + 3 D sphere.

Retinoscopy is usually performed in a dark room. The examiner sits at a distance of 1 meter from the patient and the latter wears a trial frame. A light source is placed above and behind the patient's head. The surgeon reflects the light by a plane mirror into the patient's eye and then the mirror is slowly tilted from side-to-side horizontally and then vertically. The direction in which the red reflex moves is noted. In high degree of ametropia, the reflex has a curved border, is dull and moves slowly. But in low refractive error, the reflex is bright, with straight border and moves rapidly. As the periphery of the cornea is flatter than the center, the reflex movements differ centrally and peripherally. For refraction, only the central reflex movements should be observed. In practice, the horizontal meridian is observed first. If the reflex moves with the mirror, progressively stronger convex lenses are put in the trial frame until a point of reversal is obtained. If the refraction is equal in both the meridians, only spherical correction is needed. In astigmatism (regular), the reflex movements vary in different meridians. Each principal meridian should be neutralized separately. When one meridian is neutralized, the shadow becomes band-shaped, the edge of the band being parallel to the axis of the neutralized meridian. The mirror is then moved at right angles to the neutralized meridian and a point of reversal is obtained by adding the lenses in the trial frame.

Retinoscopy in elderly patients is generally done without the use of any cycloplegic. However, a cycloplegic is essential for the estimation of refractive error in children as they have strong accommodative reserve. Hypermetropes below the age of 20 years may need homatropine hydrobromide (2%), tropicamide (1%) or cyclopentolate (1%) to relax their accommodation during retinoscopy. If there is a marked disparity between objective and subjective findings and if there are symptoms of accommodative asthenopia, refraction under cycloplegia is recommended. In such cases a correction must be made to compensate for physiological tone of the ciliary muscle (add – 0.5 to 1 D). The use of cycloplegic in elderly persons with shallow anterior chamber is contraindicated because it can precipitate an attack of acute congestive glaucoma.

Retinoscopy for near vision is not done in practice for all cases. An objective measurement of the state of refraction of the eye when focused for near vision is known as *dynamic retinoscopy*. Generally, near correction for a presbyope is advised over a distant correction considering the patient's age, accommodative ability and working distance.

KERATOMETRY (OPHTHALMOMETRY)

Keratometry is a technique which measures the curvature of the cornea with the help of an ophthalmometer or a keratometer (Fig. 7.4). It is especially useful in the assessment of corneal astigmatism. Since some amount of astigmatism may occur due to lenticular factors, the technique is not reliable in determining the total astigmatic error of a patient except in aphakia. Keratometry is based on the fact that the front surface of the cornea acts as a convex mirror and the size of the image of an object reflected by it varies inversely with its curvature. The instrument consists of two illuminated 'mires' mounted on a rotatable circular arc, and an attached telescope (Fig. 7.5A). The mires are reflected on the cornea of the patient and one observes four images of the mires through the telescope. The two peripheral images are

Fig. 7.4: Keratometer (Bausch and Lomb type model)

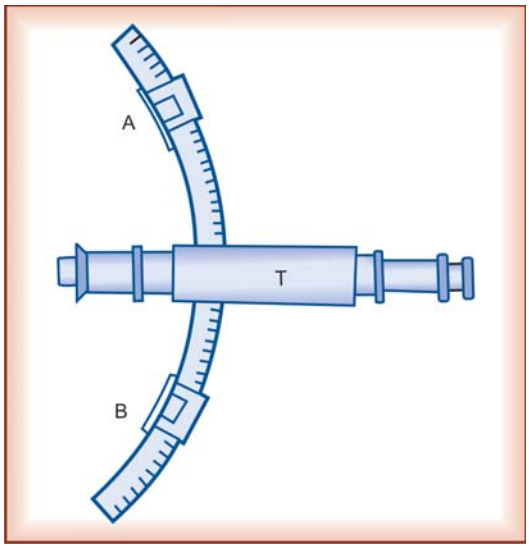


Fig. 7.5A: A and B: illuminated mires in Javal and Schiotz keratometer, T: telescope

discarded and the remaining two are adjusted in such a way that they coincide with each other at their inner edges (Fig. 7.5B). The radius of curvature and refractive power can be read from a graduated scale attached to the arc. The arc is now rotated through 90° and if the images of the mires

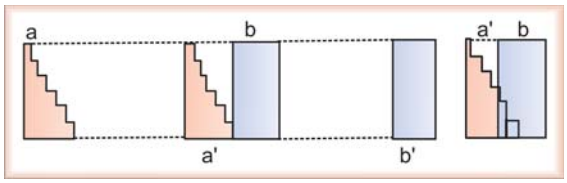


Fig. 7.5B: ab: mires, a'b': duplicate images of the mires on the cornea, a'b: overlapping of the mires when the curvature is greater

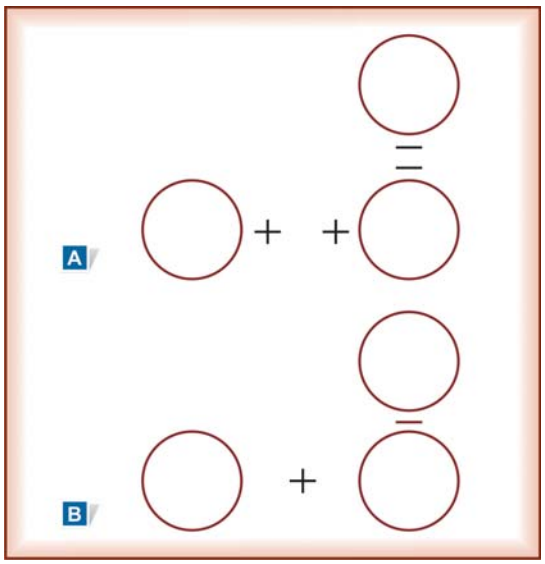


Fig. 7.6: Bausch and Lomb keratometry mires. A: Non-aligned mires, B: Aligned mires

remain unchanged, there is no astigmatism. In the presence of astigmatism, the mires will overlap or separate, hence, readjustment is required.

Generally, the mire is so constructed that each step corresponds to 1 D of astigmatism. The mires appear grossly distorted in irregular astigmatism and no useful reading can be obtained.

Baush and Lomb keratometer measures both refractive power (in diopters) and radius of curvature (in mm) of the cornea. The instrument has 2 maneuverable prisms aligned vertically and horizontally. In addition to an original image there are 2 adjustable images, one above and one to the left (Fig. 7.6). The adjustable images are moved towards or away from the original by changing the distance between the eyepiece and the prism.

SUBJECTIVE VERIFICATION OF REFRACTION

After the objective determination of refraction, each case should always be verified subjectively by testing the visual acuity and then only should the final glasses be prescribed. The post-mydriatic test should be delayed for 2 weeks when atropine is used, for 48-72 hours if homatropine or cyclopentolate is applied and for a day following tropicamide-induced cycloplegia so that the physiological ciliary tone is restored. An exception is made in cases of infants and young children for whom glasses are prescribed after making allowances for cycloplegia and working distance from the retinoscopic findings. A trial frame (Fig. 7.7) is put on the face of the patient. As a general rule, the weakest concave lens or the strongest convex lens (in myopia and hypermetropia, respectively) from the trial case (Fig. 7.8) is placed in the trial frame using an occluder for the fellow eye. Now the power of the lenses is gradually increased or decreased until maximum visual acuity (6/6) is obtained by using Snellen's distant test-types (Fig. 7.9). The same procedure is repeated for the occluded eye and finally the acceptance is verified binocularly.

Fig. 7.7: Trial frame

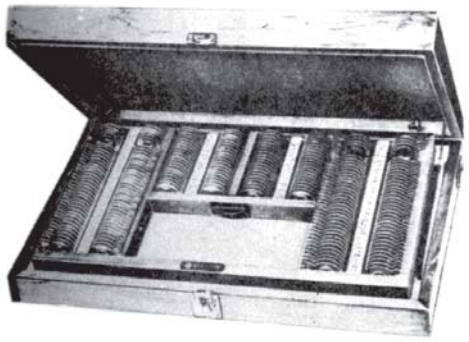


Fig. 7.8: Trial case

Fig. 7.9: Distant vision drum—wall model

Some surgeons apply *fogging method* to relax the ciliary muscle. In this method, the patient is made myopic by 1 D by addition or subtraction from the retinoscopic findings. Then concave spherical lenses (in 0.25 D steps) are gradually added until maximum visual acuity is obtained. If the vision does not improve to 6/6, cylindrical lenses should be tried as per the retinoscopy. The axis of cylinder should be rotated a few degrees

Fig. 7.10: Maddox astigmatic fan (*Courtesy:* Punjab Surgicals)

Fig. 7.11: Cross-cylinder

on either side to see whether there occurs any further improvement in vision.

The degree and axis of astigmatism can be determined by the use of either an astigmatic fan (Fig. 7.10) or a cross-cylinder. The *astigmatic fan* is made up of radiating black lines in different meridians separated by 10° interval. If the patient is stigmatic, all the lines appear equally clear to him. But if there is astigmatism, he sees some of the lines more clearly than the others. The test is carried out after fogging the patient's vision by adding + 1 D to the trial lens. The patient is asked to look at the fan. In the presence of astigmatism, the patient will see some of the lines more sharply defined. Concave cylinder is now added with its axis at right angles to the clearest line until all the lines appear equally clear. The exact axis of the cylinder can be verified with the help of a rotating 'V' in the center of the fan. When both the arms of 'V' appear equally clear to the patient, the apex of 'V' coincides with the more ametropic meridian. The axis of the cylindrical lens should lie at right angles to this meridian.

A *cross-cylinder* is a combination of two equal cylinders of opposite signs with their axes at right angles (Fig. 7.11). The most popular combination is + 0.25 D. It is used for subjective refinement of the axis and the power of the prescribed cylinder. Jackson cross-cylinder enlarges or contracts the interval of Sturm, blurring or clarifying the image formed on the retina, by increasing or decreasing the astigmatic ametropia.

To check the strength of the cylinder in the optical correction, the axis of the cross-cylinder is first placed in the same direction as to the axis of the cylinder in the trial frame and then perpendicular to it. If in both the instances visual acuity remains unchanged, the cylinder in the trial frame is correct. Should the visual acuity change, a suitable alteration in the strength of the cylinder is to be made.

The axis of the correcting cylinder can be verified by placing the cross-cylinder obliquely (at 45°) to the axis of the correcting cylinder in the trial frame. Should the patient read the test types more clearly, the axis of the cylinder in trial frame be rotated till the letters appear equally clear and rotation of the cross-cylinder gives no alteration in distinctness.

The over or under optical correction can be verified by *duochrome test*. It is based on the phenomenon of chromatic aberration. The green rays, having a short wavelength, are refracted more acutely and brought to a focus earlier than the long red rays. When ametropia is fully corrected, the eye becomes emmetropic and a focus is formed between these two extremes. If it is myopic, red is seen more distinctly, whereas a hypermetrope sees green more sharply. The test is carried out by asking the patient to read letters over the colored panels on the vision drum. Both green and red letters should appear equally blurred by optical correction. If the red letters are clearer than the green in myopia, it is certain that over-correction has not been done. But if the patient sees green letters more distinctly than the red, the patient is over-corrected. Each eye should be separately tested to rule out the over or under-correction.

In all conditions where the visual acuity does not improve with the optical correction, a *pin-hole disk test* should be performed. An occluder disk with a central hole of 0.50 to 1.5 mm diameter (Fig. 7.12) is placed in the trial frame. If the visual acuity improves, the refraction should be rechecked. However, no improvement in the visual acuity even with the pin-hole disk indicates some organic lesion in the macula.

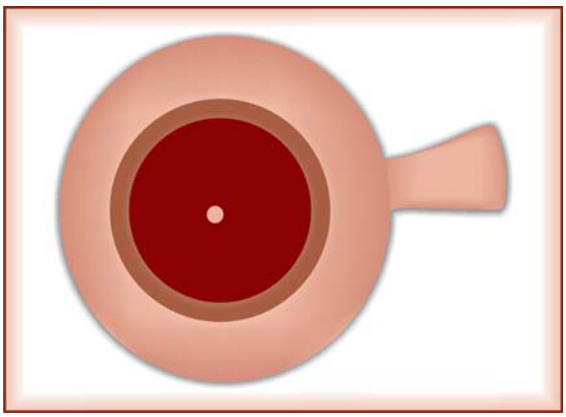
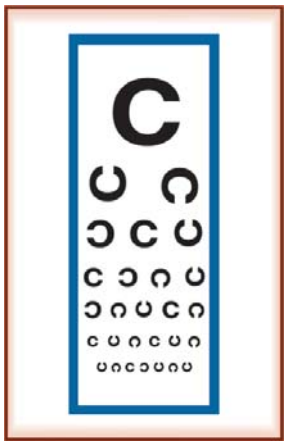


Fig. 7.12: Pin-hole disk

CORRECTION OF NEAR VISION

The correction of near vision should be preceded by the distant correction and determination of near point with the distant correction in place. Ordinary types used in printing are utilized for the correction of near vision (Fig. 7.13). Jaeger's types, Number point types standardized by the faculty of ophthalmologists (N 5 to N 45) and Snellen's reading test-types or broken C may also be used. The patient is asked to hold the test-types at a distance at which he is accustomed to read or



N 6

When I was ten years old, my father had a small estate near Satara where he used to take us during the holidays. It was situated in rough and uncultivated countryside where wild animals were often seen. Once we heard that there was a panther in the surroundings who was killing the cattle and attacking the villagers. Father had warned me not to wander far from home in the evenings. I had made friends with a young villager called Ramu.

N 8

The cattle were slowly making their way home in front of us. The dog which helped Ramu ran barking at the hooves of the cows, who sometimes made a playful rush at the dog. Crows and mynas in flocks were passing home over our heads.

N 12

The cow was knocked over and I saw the tiger sitting over its white body. The cow kicked and struggled.

Fig. 7.13: Number point types for near vision testing

work. If the letters are not distinctly seen, suitable spherical convex lenses are added to the distant correction so that the types are easily and comfortably read. Now the near point should be determined. This is done by gradually bringing towards the eye a card on which is drawn a line 0.2 mm in width until the line appears blurred. It can also be measured by using the smallest test-types; moving it gradually towards the eye until they appear blurred or no longer be easily read. The distance of the near point from the eye can be measured with a tape. The range of accommodation can be calculated from the formula A = P – R (*vide infra*). The near correction given should be such that nearly 1/3 of the amplitude of accommodation is kept in reserve. Generally, it is better to under-correct than to over-correct because stronger convex lenses may cause difficulties in convergence and the range of near vision will be limited. In cases where strong correction is needed on occupational ground, the incorporation of prism facilitates convergence.

PRESCRIPTION OF SPECTACLES

The prescription of spectacles should have relevance with the patient's symptoms. When refractive error is associated with visual defect, there is an obvious indication for the prescription of the spectacles. Spectacles should not be used as placebo therapy unless ocular symptoms are predominant. Prescription of spectacles for slight degrees of hypermetropia is better avoided.

The refraction of young children with myopia or hypermetropia needs periodical check ups, a six monthly review is often helpful. Most adult subjects have static refraction. Scratched lenses or worn out frames warrant a change.

Types and Fitting of Spectacles

The spectacle lenses are made of crown glass or of hard thermosetting resin (allyl diglycol carbonate monomer). The *resilens* or CR-39 plastic lenses are light and resistant to scratching. They can be dyed to reduce the transmission of light and surface coated to check glare (*anti-reflective coating*). The lenses should be securely fitted in light, strong and rigid frames. The design of the frame should be such that the rim's bridge, side pieces and joints should not press the patient's nose or temple. The lenses be held at a distance of about 15 mm in front of the cornea corresponding to the anterior principal focus of the eye as at this distance the images formed on the retina are of the same sizes as in emmetropia.

The *high-index lenses* have the ability to bend light rays more as compared to normal lenses. They have refractive index ranging from 1.53 to 1.74. High-index plastic lenses are commonly ordered for high myopes as they are thin, light in weight and cosmetically more acceptable.

Polycarbonate material is used for lenses and frames in children and sportsmen. It is scratch and impact resistant. *Polycarbonate lenses* are thin, light in weight and have inherent property of ultraviolet protection.

There is a gradual change in the curve of front surface of the lens from center to periphery in *aspheric lenses*. Therefore, they do not cause spherical aberration. The aspheric lenses are extremely useful in high degrees of myopia and hypermetropia. Besides having superior optics, they are thinner and lighter. The thinnest edge for a strong minus power is produced when the aspheric lens is made of a high-index material. Moreover, the cosmesis in patients of high myopia can be further improved by special edge polishing and buffing, and mounting these lenses on plastic (cellulose acetate or zylonite) frames.

It is important that the wearer must view through the spectacles and not be tempted to look over them. Therefore, in children large glasses are provided. In astigmatism, the use of oval glasses prevents rotation should the frame become loose.

Rigid spectacles are ordered for adult astigmatic patients. Every care should be taken to center the lenses so that their optical centers lie opposite the center of the pupil. The presbyopic lenses are slightly decentered inwards as the eyes are directed downward and inward in reading.

The *bifocal lenses* are quite popular in presbyopia wherein the upper segment contains the distant correction and the lower the near correction. In *trifocal lenses*, a strip of an intermediate distance is interposed between the distant and the near correction. Multifocal lenses are also known as *progressive* or *gradient lenses* in which a continuous gradation from the near to the far point is incorporated. The progressive or no-line bifocal lenses provide a smooth transition from distant through intermediate to near vision. There are certain inherent disadvantages of progressive lenses such as difficulty in moving, particularly going downstairs, blurring and prismatic effects.

Tinted glasses are advised for albinism, high myopia and glare-prone patients. *Photochromic lenses* are also popular. They contain ultraviolet activated color changing silver halide molecule that makes the lens change from light to dark on exposure to sunlight.

CONTACT LENSES

The contact lens is worn in apposition with the cornea (Fig. 7.14). The quality of the image viewed through contact lenses is far superior than that seen through the spectacle lenses.

The contact lenses are of three types: (i) hard lenses, (ii) gas permeable lenses, and (iii) soft lenses.

Hard Contact Lenses

The hard lenses have poor oxygen permeability. However, they can correct a high corneal astigmatism as in keratoconus. The hard contact lenses are made up of polymethylmethacrylate (PMMA) and may be scleral or corneal.

1. *Scleral (haptic) lenses* cover the cornea and rest on the sclera.
2. *Corneal lenses* cover the cornea and have a diameter less than that of the cornea.

Gas Permeable Contact Lenses

Rigid gas permeable (RGP) contact lenses are made up of copolymer of PMMA and silicone

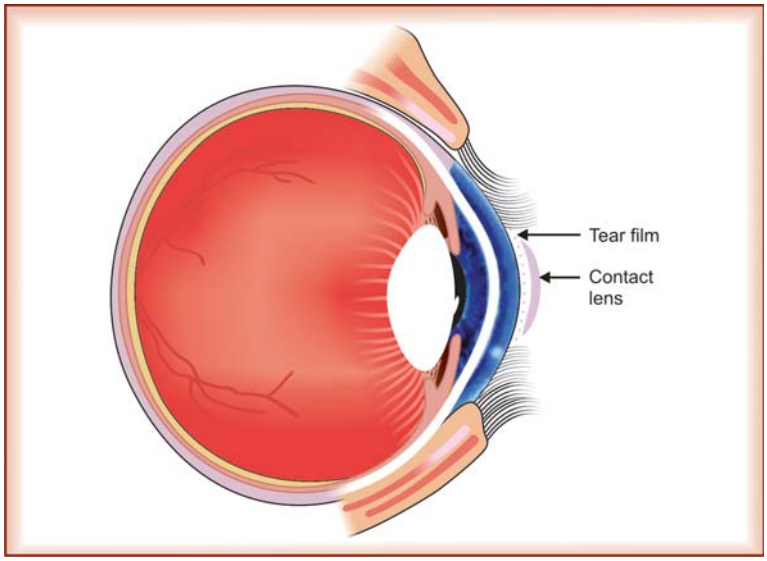


Fig. 7.14: Contact lens

containing vinyl monomer. The lenses are softer than PMMA lenses and permeable to oxygen. They have a broader optical zone than hard lenses and they provide good vision both in day and night. Gas permeable lenses are more comfortable and are less likely to pop out of the eye than the hard lenses. Owing to their relative softness they get more easily damaged.

Soft Lenses

Soft contact lenses are more comfortable and well tolerated than the hard and gas permeable lenses. They are made up of hydroxyethylmethacrylate (HEMA). The soft lenses just overlap the limbus and are oxygen permeable. The visual acuity of the patient wearing soft lenses fluctuates with the blinking. Soft lenses have short life span and have relatively poor optical quality than the hard lenses. They are prone to protein deposits and may be contaminated by microorganisms. A soft contact lens seldom corrects astigmatism of more than 1D but the use of *toric soft contact lens* can satisfactorily improve the vision even in high astigmatic patient.

Extended wear soft contact lenses are made from highly oxygen permeable silicone hydrogel material. They can be worn continuously for 7 to 30 days without removal, cleaning or disinfection. The lenses that are used for a specific period of time, then disposed off and replaced with a new pair of lenses, are known as *disposable contact lenses*. They can be daily, weekly or monthly disposables. Substances like protein, lipids and calcium, found in normal tear-film, can get deposited on the contact lenses making their replacement mandatory.

Special Types of Contact Lenses

1. *Therapeutic contact lenses*: Ultrathin contact lenses are used as *bandage contact lenses* in patients with corneal erosions and for topical drug-delivery.
2. *Iris-print contact lenses*: Iris-print lenses, with an opaque iris-print and a clear pupil, may be used in patients with albinism and aniridia. They reduce glare and provide good cosmetic effect.
3. *Bifocal contact lenses*: These contact lenses are indicated in patients wih presbyopia.

Merits and demerits of different types of contact lenses are listed in Table 7.1.

Optics of Contact Lens

The contact lens reduces the cornea to an insignificant optical surface due to its close adherence to the tear film. Thus, the contact lens imparts its optical correction mainly by changing the power of the eye. If the cornea is scarred and has irregular astigmatism, the error is eliminated by the tear film lying between the cornea and the contact lens.

Table 7.1: Merits and demerits of different types of contact lenses

Property	*Hard*	*Soft*	*RGP*
Visual acuity	Good	Affected by blinking	Clear
Use in astigmatism	Possible	Less suitable	Possible
Oxygen delivery	Poor	High	Moderate
Adaptability	Needed	Not needed	Needed
Deposits	A few	Considerable	A few
Durability	Undergo scratching	May undergo tear	Scratch and tear not common

Indications

Contact lenses are indicated for following purposes.

1. Optical
2. Cosmetic
3. Occupational
4. Therapeutic, and
5. Preventive.

Optical indications include anisometropia, unilateral aphakia, high myopia and irregular astigmatism (keratoconus and corneal scar).

Cosmetic indication includes unsightly and disfigured eye.

Occupational contact lenses are worn by stage artists, sportsmen and pilots.

Therapeutic indications include corneal epithelial healing defects, recurrent corneal erosions, bullous keratopathy and wound leak. The contact lens is also used as a vehicle for drug delivery.

Preventive indications include exposure keratitis, trichiasis and prevention of symblepharon in cases of membranous conjunctivitis and chemical burns.

Advantages

The contact lens has several advantages over spectacles. The use of contact lenses can retain binocularity in anisometropia owing to less magnification of the size of the retinal image. The contact lens moves with the eye and eliminates the peripheral distortion caused on eccentric viewing through a powerful spectacle lens. Contact lens is not subjected to moistening up. It is cosmetically superior. Tinted contact lenses relieve photophobia in cases of albinism. Colored contact lenses are worn to change the color of eye temporarily, and for special eye effects—*costume* or *theatrical lenses*.

Disadvantages

Contact lenses are not without disadvantages. A poorly fitting contact lens may cause corneal edema or abrasion. Most of the corneal complications of contact lens occur due to the deprivation of oxygen to the cornea. The lenses cannot be worn comfortably in a dusty and dirty environment. They are capable of inducing an allergic reaction in the conjunctiva. *Giant papillary conjunctivitis* (GPC) is seen in some patients with hydrogel contact lenses. Thimerosal, a preservative found in many contact lens solutions, is also responsible for GPC. Soft lenses are prone to carry infections. The complications are more common with the extended wear contact lenses.

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CHAPTER

8 Accommodation and its Anomalies

ACCOMMODATION

The parallel rays coming from infinity are focused on the retina in emmetropic eyes. But the light rays from an object within 6 meters are divergent; they form a focus behind the retina. The normal eye is capable of seeing the near objects clearly due to an increase in the power of the lens which is brought about by the increased convexity of the anterior surface of the lens. This ability of the eye to change its refractive power is known as *accommodation*.

The mechanism of accommodation is regulated by an accommodational reflex which is induced by the blurred images of the near objects. The visual impulses relayed from the cortex reach the Edinger-Westphal nucleus of oculomotor nerve causing contraction of the ciliary muscles resulting in forward movement of the external surface of the ciliary body. It leads to the relaxation of the suspensory ligament of the lens which allows the anterior surface of the lens to become more convex. According to Helmholtz the lens becomes smaller and thicker during accommodation. Fincham postulated that the shape of the lens is determined by the structure of the capsule. The central zone of the anterior surface becomes more convex in relation to the peripheral part of the lens. The posterior surface of the lens undergoes little change in curvature as it is well supported by the anterior face of the vitreous. The radius of curvature of the anterior surface of the lens at rest is 10 mm and that of the posterior surface 6 mm. During strong accommodation the radius of curvature of the anterior surface becomes 6 mm, while no change occurs in the radius of curvature of the posterior surface (Fig. 8.1).

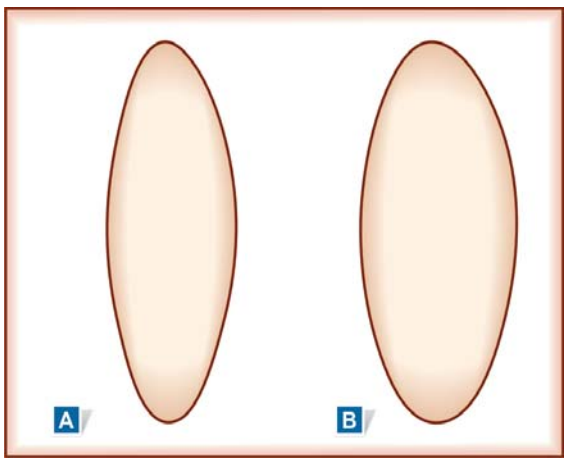

Fig. 8.1: (A) Anterior surface of an unaccommodated lens (B) Anterior surface of an accommodated lens

The near point (*punctum proximum*) and the far point (*punctum remotum*) of the eye vary with age and the state of refraction. The difference between the refractive power of the eye at the near point (when the accommodation is maximum) and at the far point is called the *amplitude of accommodation*.

The amplitude of accommodation (A) is expressed by the formula, A = P – R, wherein, P represents the refractive power of a fully accommodated eye (i.e. reciprocal of the distance of the near point in meters) and R is the refractive power of the eye at rest (i.e. reciprocal of the distance of the far point in meters).

Amplitude of accommodation is a monocular expression of change in lens power in diopters and is measured either by measuring the near point of accommodation using small prints or with the help of an accommodation rule (fixation rule).

The *near point of accommodation* can be measured by using a fixed print size type and moving the type towards the eye until the print blurs. The point of blur measured in centimeters to the spectacle frame is recorded (Donder's push-up method). An emmetropic eye has its far point at infinity and the measured near point can be converted into diopters of amplitude of accommodation (100/Point of blur in cm).

Krimsky Prince near point accommodation rule consists of a reading card with a ruler calibrated in centimeters and diopters. With this bifurcated rule, direct readings of accommodation near point and convergence near point can be made. The binocular amplitude of accommodation is usually greater than monocular by 0.5 to 1 D.

Range of accommodation is defined as the distance between the far point of the eye and the near point at which the eye can maintain a clear vision. The measurement of range of accommodation is of practical importance while giving the presbyopic correction. It can be done with the help of an ordinary tape, scale or accommodation rule. The determination of range is related to the diopteric power required by an individual to perform a specific job.

The amplitude of accommodation varies inversely with age. It diminishes as age advances. The average amplitude of accommodation is about 14 D (+ 2 D) at the age of 8 years, and it gradually diminishes to 10 D at the age of 24 years, 6 D at 40 years, 3 D at 48 years, 2 D at 56 years and 1 D at 64 years. A rough estimate indicates that there occurs a rapid decrease in the amplitude of accommodation between the age of 40 and 48 years.

The accommodation also varies with the state of refraction of the eye. In an emmetropic eye, the far point lies at infinity and for distant vision, therefore, the eye is at rest. If the near point of an emmetropic eye lies 10 cm away, then P = 100/10 = 10 D. The range of accommodation is, therefore, infinity to 10 cm. On the other hand, a hypermetrope in order to see a distant object exerts an amount of accommodation equivalent to his hypermetropia, and to see an object at 10 cm further accommodates by 10 D to put him at par with an emmetrope. Thus, the amplitude of accommodation of a hypermetrope is greater than an emmetrope. A hypermetrope makes a continuous use of his accommodation both for distant and near works and the demand made upon the ciliary muscle is much more for the near work. A myope has the far point in front of his eyes. He cannot see the distant object clearly by any effort of accommodation but can see the near object with less effort than an emmetrope or a hypermetrope. The accommodative effort cannot counteract an astigmatic error, hence, a distinct image is never obtained in astigmatism. As the accommodative effort of the two eyes cannot be dissociated, it cannot correct anisometropia.

Besides the increase in the curvature of the anterior surface of the lens, convergence of the eyes and constriction of the pupils occur during accommodation. There is a close relationship between accommodation and convergence, and normally one meter angle of convergence is found with one diopter of accommodation. The constriction of the pupil followed by accommodation and convergence is a synkinetic action which increases the depth of focus and minimizes spherical aberration.

ANOMALIES OF ACCOMMODATION

Anomalies of accommodation are not uncommon. They can be classified as follows:

1. Presbyopia
2. Insufficiency of accommodation
3. Paralysis of accommodation, and
4. Spasm of accommodation.

Presbyopia

Presbyopia is not a refractive error but a physiological condition of gradual loss of accommodative power due to age-related decrease in the elasticity of lens capsule and lens substance. Besides lenticular changes, loss of ciliary muscle function is also implicated in the development of presbyopia.

Clinical Features

For comfortable near work the amplitude of accommodation must be double the amount of accommodation an individual needs. The symptoms of presbyopia usually begin near the age of 40 years with a decline in a person's amplitude of accommodation. The onset may be heralded by development of asthenopic symptoms with blurring of near vision especially in dim light, heaviness of eyes, or tiring of eyes on prolonged near work. The patient prefers to keep the near objects, newspapers and books at a greater distance than usual because less accommodative effort is needed at an increased distance. The onset of presbyopia will also depend on the refractive error (early in hypermetropes and late in myopes), the depth of focus and the nature of visual task.

Treatment

The goal of correction of presbyopia is to strengthen the amplitude of accommodation by prescribing the convex lenses for near vision. Initially the refractive error for distance is determined and corrected. Then the patient is asked to read the near test-types at the distance he/she commonly works at. The weakest plus (convex) lens required to read the smallest type is added; it is generally + 0.75 D to + 1.25 D at the age of 40 years. As the age increases, the amplitude of accommodation decreases and the power of convex lens increases. However, no attempt should be made to overcorrect presbyopia, otherwise the patient has to work at a close distance resulting in discomfort.

Presbyopic spectacles can either be single vision reading glasses or bifocals in which an additional plus lens is added to the lower portion of a distant vision lens.

Presbyopia can also be managed surgically. The surgical correction is achieved by presby-LASIK which aims for obtaining corneal multifocality, intraocular phakic multifocal lenses, intracorneal lenses or intraocular pseudophakic multifocal lenses.

Insufficiency of Accommodation

When the accommodative power is below the lower limit of the accepted normality for the patient's age, it is called *insufficiency of accommodation*. The insufficiency of accommodation occurs either due to early onset of presbyopia owing to lenticular sclerosis or because of weakness of ciliary muscles as found in anemia, toxemia of pregnancy, malnutrition and glaucoma.

The patient suffers from eyestrain, particularly during near work. The condition can be corrected by prescribing the weakest convex lens which facilitates near work and stimulates the accommodation. The general condition of the patient should be improved.

Paralysis of Accommodation

The paralysis of accommodation or *cycloplegia* occurs as a complication of diphtheria, trauma,

syphilis, meningoencephalitis, diabetes and alcoholism or it can be induced by application of a cycloplegic drug such as atropine. Paralysis of accommodation is often accompanied by dilatation of pupil (mydriasis) owing to complete paralysis of sphincter pupillae (excepting diphtheria).

Photophobia and blurred vision are the common symptoms. Recovery is seen in drug-induced paralysis and in cases of diphtheria. The prognosis is also good in traumatic cycloplegia. Dark glasses and suitable convex lenses for near work are prescribed. Instillation of miotics is seldom beneficial.

Spasm of Accommodation

The spasm of accommodation or *cyclotonia* may be found in young myopes engaged in excessive near work in poor illumination. It can be produced artificially by instillation of strong miotics. Generally, ciliary muscle has a physiological tone of about 1 D, but in spasm of accommodation this tone becomes much greater. It can be revealed by application of atropine.

The patient has a refractive error, usually myopia of relatively high degree on subjective testing (*pseudomyopia*). Refraction under atropine indicates the actual error which should be carefully corrected. Near work must be curtailed and cycloplegic drop is prescribed for sometime to relax the spasm.

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CHAPTER

9 Examination of the Eye

HISTORY-TAKING

It is not only customary but essential to record the complaints of the patient before starting the actual examination of the eye. Like other branches of medicine, the eye patients should be encouraged to describe their ailments. A proper record of history should be maintained.

Defective vision (for distance, near or both), discharge from the eye, redness, photophobia, itching, burning or foreign body sensation and ocular pain or discomfort associated with dull or severe headache are some of the common complaints of the eye patient. The mode of onset (acute or insidious) and duration of the ailment should be enquired. The nature of the discharge—watery, mucopurulent, purulent, sanguineous or ropy—must be verified. The association of itching and burning of eye with the change in season or climate should be looked into. The severity of the ocular pain and its relation with close work or systemic disorders like hypertension or migraine should be ascertained. Any history of trauma, blunt or penetrating, or retained foreign body is taken because such cases may need emergency intervention.

The age of the patient is an important factor in visual disability. Senile cataract and glaucoma predominantly affect a person after fifth or sixth decade. The near vision of an otherwise normal individual suffers a setback in presbyopic period (above 40 years). Youngsters show an increase in the rate of progression of myopia, particularly at puberty.

Refractive errors are seen in the patients of all age groups. However, they produce discomfort in persons engaged in accountancy or fine precision work. Industrial workers are exposed to occupational hazards and some of them may report with serious injuries to the eyes.

The patient should be asked about coexisting systemic illness and ongoing treatment. A probe into the past medical and surgical history of the patient provides important clues in establishing the correct diagnosis of the present illness.

A family history is helpful in confirming the inheritable ocular disorders like ptosis, squint, glaucoma, dystrophies, etc. Questioning may be done regarding any fever during the first trimester of pregnancy (rubella), venereal diseases and application of forceps at the time of the delivery, as they often cause ocular anomalies.

EXAMINATION OF THE EYE

The examination of the eye in children requires the help of an attendant who can wrap the child in a piece of cloth and holds the child on his/her legs, the baby's head being fixed between the surgeon's knees. For the proper visualization of the cornea and the anterior chamber, lid retractor must be used. An adequate magnification either by a monocular loupe (Fig. 9.1) or a binocular

loupe (Fig. 9.2) or by a slit-lamp biomicroscope (Fig. 9.3) with brilliant illumination helps in the evaluation of the external eye and the anterior segment of the eye. If the child is uncooperative or irritable examination under sedation or general anesthesia is recommended.

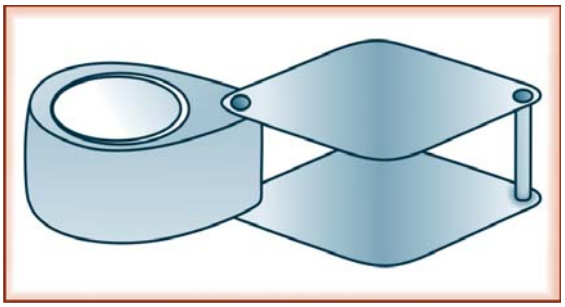


Fig. 9.1: Corneal loupe

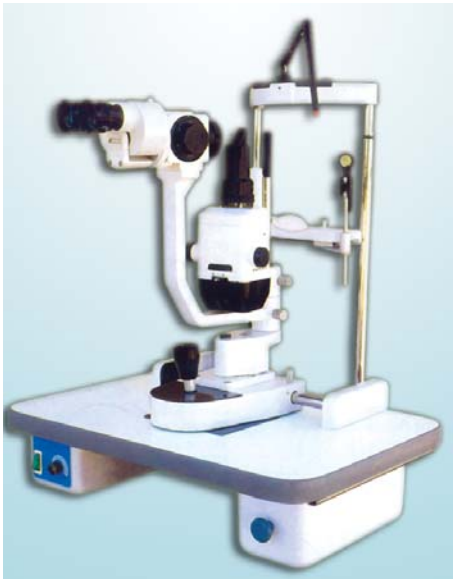


Fig. 9.3: Slit-lamp

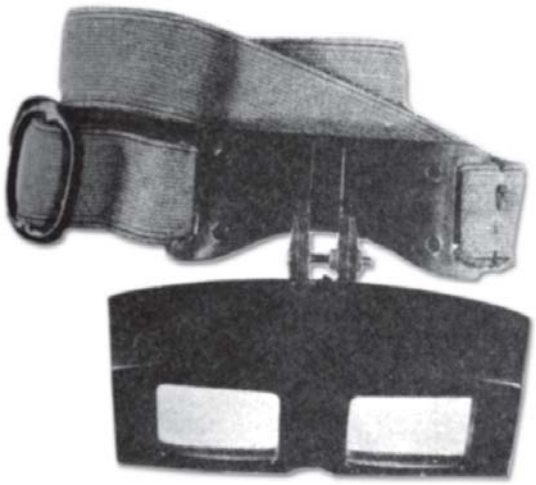


Fig. 9.2: Binocular loupe

A complete examination of the eye includes recording of visual acuity, color vision, field of vision, examination of ocular appendages (eyebrows, lids, lacrimal apparatus and conjunctiva), anterior segment of the eye (cornea, anterior chamber, angle of the anterior chamber, iris, pupil and lens) and posterior segment of the eye (vitreous, retina, choroid and optic nerve). The examination should be carried out in day-light or in a well-illuminated room.

Before commencing the eye examination, the position of the head and chin may be noted as the patient of strabismus, particularly in case of vertical muscle palsies, often keeps the head tilted and chin elevated to avoid diplopia.

The face must be inspected for any asymmetry which is common following Bell's palsy or in cases of facial hemiatrophy or facial muscular dystrophy. The forehead may show excessive wrinkling, a sign of frontalis overaction, to compensate the underaction of levator palpebrae superioris in partial ptosis. On the other hand, complete loss of wrinkling on one half of the forehead denotes lower motor neurone facial palsy. The presence of unilateral pitted scars above the eyebrow and on ipsilateral side of the nose suggests an attack of herpes zoster ophthalmicus in the past. The eyebrows may show scanty hair, especially in leprotic or myxedemic patients.

Normally, both the eyeballs are symmetrical and so placed in the orbital cavities that the

anterior convexities of the eyes do not extend more than 12 to 20 mm from the summit of the lateral orbital margins. Sometimes, one or both eyeballs may bulge beyond this limit giving rise to what is known as *proptosis* or *exophthalmos*. On the contrary, the eyeball may be deeply set in the orbit, *enophthalmos*.

The palpebral fissure, the exposed space between the margins of two lids, in adults measures 28-30 mm in length and 10-14 mm in width. However, in inflammatory conditions of the conjunctiva and cornea, due to blepharospasm, it remains narrow. The level of medial and lateral canthi is more or less same, but in Mongolians the lateral canthus is at a higher level than the medial leading to an obliquity of the aperture. This peculiar shape is called as *mongoloid obliquity of the palpebral aperture*. Inversely, one can find an *antimongoloid obliquity of the palpebral aperture* in Crouzon's disease wherein the outer canthus is at a considerably lower level than the medial. The palpebral aperture may be all around narrow since birth—*blepharophimosis*. A fold of skin may run from the upper lid over the medial canthus (*epicanthus*) which is a racial characteristic of Mongolians. If this fold is prominent it can result in pseudostrabismus.

Usually the upper lid covers the upper one-sixth of the cornea. However, occasionally the upper limbus is visible due to retraction of the lid, a feature of thyrotoxicosis or sympathetic overactivity, or in proptosis, a forward bulging of the eyeball.

The eyelashes are directed forwards and laterally. An inward misdirection of a solitary eyelash causes foreign body sensation and watering. Hence, a careful inspection of the lid margin is important. A complete inrolling (*entropion*) of the lid margin is easy to diagnose. A mild sagging of the lower lid margin (*ectropion*) is commonly seen in old age and induces annoying epiphora owing to the loss of contact of the lower punctum with the lacus lacrimalis (bulbar conjunctiva). The lower lid margin just touches the lower limbus normally. An exposure of the lower limbus indicates proptosis with a pathology probably lying either in the maxillary antrum or in the orbit.

Both the eyes work in unison and during their movements their visual axes continue to maintain the alignment which can be tested by observing the corneal reflex by a torch-light. The deviation from this position results in *strabismus* which may either be comitant *(nonparalytic)* or incomitant (*paralytic).*

When both eyes show rhythmic oscillations, it is called *nystagmus*, a sign which indicates that fixation reflex is not well-developed. When the vision is impaired in infancy, the eyes often move arrhythmically or show searching movements which are known as *nystagmoid movements*.

Examination of the Conjunctiva

The lower palpebral conjunctiva is exposed by pulling down the lower eyelid (Fig. 9.4), while the patient is looking upward. However, the inspection of upper palpebral conjunctiva necessitates eversion of the upper lid which requires some practice. The patient is asked to look down and the eyelashes of the upper lid are held between the thumb and the index finger. The index finger of the other hand or a swab-stick is then placed on the upper border of the tarsal plate and the lid is rotated around the swab-stick (Figs 9.5 and 9.6).

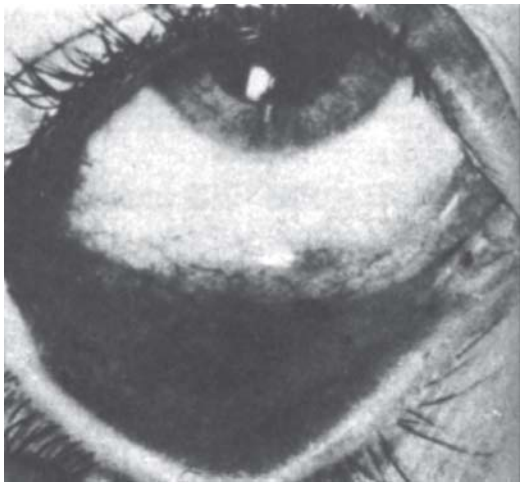


Fig. 9.4: Eversion of the lower lid

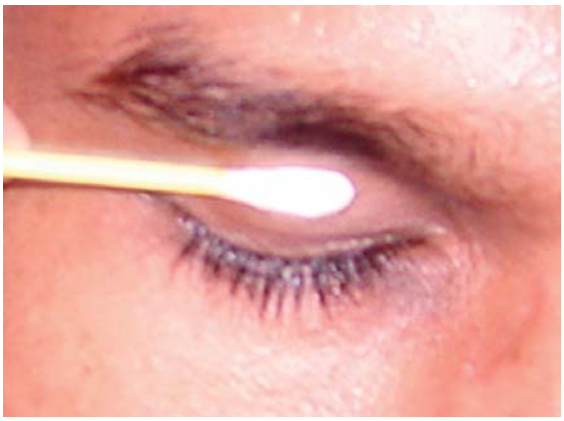


Fig. 9.5: Swab-stick placed above the upper border of tarsal plate

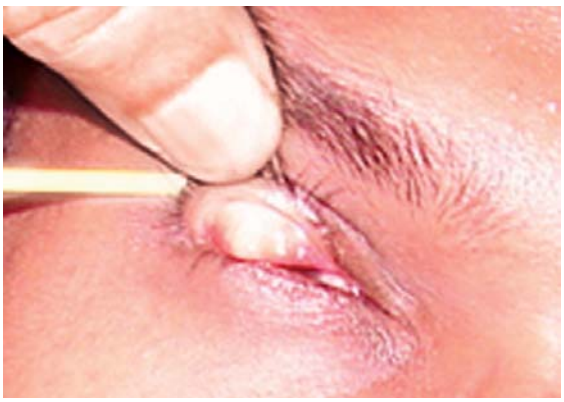


Fig. 9.6: Everted upper lid (*Courtesy*: Drs Srikant and Santosh Kumar, IMS, BHU,Varanasi)

Normal palpebral conjunctiva is translucent, smooth and presents vertically arranged thin blood vessels. It should be inspected for the presence of follicles, papillary hypertrophy, scarring, membrane, foreign body (especially in sulcus subtarsalis) and concretions. For examining the upper fornix one requires double eversion of the lid with the help of a retractor. The upper fornix is a common site for the presence of follicle, chemosis or, rarely, foreign body.

The bulbar conjunctiva can be examined by separating the upper and lower lids with fingers. The bulbar conjunctiva is a smooth, lusterful, semi-transparent membrane. Two sets of blood vessels, the posterior conjunctival vessels coming from the fornices and the anterior ciliary vessels, are distinctly seen. They join to form a fine limbal plexus. In inflammatory diseases of the conjunctiva, cornea and anterior uvea, congestion of the individual set of vessels provides a clue to the site of inflammation.

The distinction between the conjunctival and ciliary congestions should be made on the points listed in Table 9.1.

Table 9.1: Differences between conjunctival and ciliary congestions

Features	*Conjunctival congestion*	*Ciliary congestion*
1. Site and arrangement of blood vessels	Away from the limbus usually towards fornices and often branched	At the limbus and radially arranged
2. Color	Bright, brick red	Purple, dull red
3. Individual vessel	Can be seen	Cannot be distinguished
4. On moving the conjunctiva	Conjunctival vessels also move	Ciliary vessels remain stationary
5. On squeezing the blood vessel	Fills slowly from the fornix	Fills at once from the limbus

Generally, the conjunctival congestion is often accompanied with a mucopurulent or a purulent discharge, an important sign of conjunctivitis. On the other hand, ciliary congestion may be accompanied with watering and suggests a deep seated inflammation of the anterior uvea, sclera or cornea, and is usually associated with dull or severe ocular pain. Occasionally, both types of congestions may co-exist as seen in acute congestive glaucoma.

Sometimes, a wing-shaped encroachment of bulbar conjunctiva over the cornea (*pterygium*) is seen. A dry, lusterless, triangular spot (*Bitot's spot*) with the base towards the limbus is seen frequently in children with vitamin A deficiency.

Examination of the Lacrimal Apparatus

The lacrimal gland is situated in the upper and outer quadrant of the orbit and the palpebral part of the lacrimal gland can be visualized by asking the patient to look inferonasally. The lacrimal gland becomes enlarged in inflammatory and neoplastic conditions. The latter often causes proptosis and downward and inward dislodgement of the eyeball.

A thorough examination of a patient having watering should be conducted to locate the site of obstruction in the tear drainage system. Size, shape and site of the lower punctum should be confirmed. Foreign bodies, especially eyelashes and concretions, can obstruct the punctum. Rarely, a stricture may develop in the canaliculus.

The lacrimal sac lies in the lacrimal fossa and it does not cause any prominence. However, it becomes swollen in acute dacryocystitis and the overlying skin becomes red and tender. A painless swelling over the sac area is suggestive of mucocele of the sac which on pressure leads to regurgitation of muco-pus from the lower punctum. A small oozing sinus on the skin over the sac is a sequel to ill-managed acute dacryocystitis. A hard painless swelling of the lacrimal sac gives suspicion of malignancy.

The most common site of obstruction in the lacrimal passage is at the junction of sac with the nasolacrimal duct which can be demonstrated on retrograde dacryocystography (radiological visualization of the lacrimal passage after injection of a radio-opaque dye).

Syringing

The syringing of the lacrimal passage to test its patency is a much simpler and commonly used procedure (Fig. 9.7). It is done after dilatation of the lower punctum by a punctum dilator. A lacrimal cannula attached to a 5 ml syringe filled with normal saline is introduced into the lower punctum. If there is a block in the lower canaliculus or at the junction of common canaliculus with the sac, the fluid cannot be pushed into the passage. However, if the passage is blocked at the junction of the sac with the nasolacrimal duct or at the opening of the nasolacrimal duct into the inferior meatus, the fluid regurgitates through the upper punctum. In a patent lacrimal passage, the saline passes into the nose without any resistance.

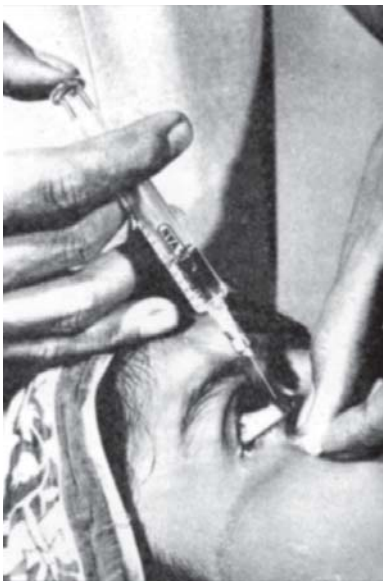


Fig. 9.7: Syringing

The patency of the passage can also be tested by either putting a drop of chloramphenicol (0.4%) or colored fluid (fluorescein 2% or mercurochrome 2%) into the conjunctival *cul-de-sac*. If the passage is patent, in the former procedure the patient will have a bitter taste and in the latter, on blowing the nose over a pad of cotton, the colored fluid will stain it.

The nasal conditions like atrophic rhinitis and polyp, can also lead to the obstruction of the opening of the nasolacrimal duct and it is, therefore, necessary to examine the nasal cavity in patients complaining of epiphora.

Primary nonsecretion (*alacrimia*) or hyposecretion of tears is an uncommon entity, while secondary dry eyes are common. Trachoma, membranous conjunctivitis, pemphigoid and ocular burns produce extensive damage to the goblet cells of the conjunctiva and strangulate the tear ducts, and thus cause *dry eye syndrome.* The tear secretion can be measured by Schirmer's test. The test is performed with standard strips of filter paper (5 × 35 mm). One end of the paper is bent and placed in the lower palpebral conjunctiva near the lateral canthus. It is left there for 5 minutes. Normal persons wet 10-30 mm of the paper, less than 10 mm of wetting indicates hyposecretion.

Examination of the Cornea

The cornea is a bright, transparent, more or less circular structure which forms the window of the eye. Even nebular changes in the corneal transparency result in visual disturbances. With a little experience, one can recognize variations in corneal diameter. Anteriorly, the cornea appears elliptical, its average vertical and horizontal diameters measure 11 mm and 11.5 mm, respectively. A small cornea, *microcornea*, that is less than 10 mm may be flat (*microcornea plana*), while a developmental increase in the corneal diameter (12.5 mm or more) causes *megalocornea* and needs to be differentiated from *buphthalmos*.

The curvature of the cornea may show a localized conical bulge (*keratoconus*), especially in young girls, or the entire cornea may appear globular (*keratoglobus*). A change in the curvature of the cornea distorts the window reflex. Placido's keratoscopic disk (disk painted with alternating black and white circles, Fig. 9.8) may be used to assess the corneal surface. On looking through a hole in the center of the disk, a uniform and sharp image of the circles can be discerned over the surface of the cornea (Fig. 9.9). But, if the corneal surface is uneven, irregularities in the rings are seen (Fig. 9.10).

Fig. 9.8: Placido's disk

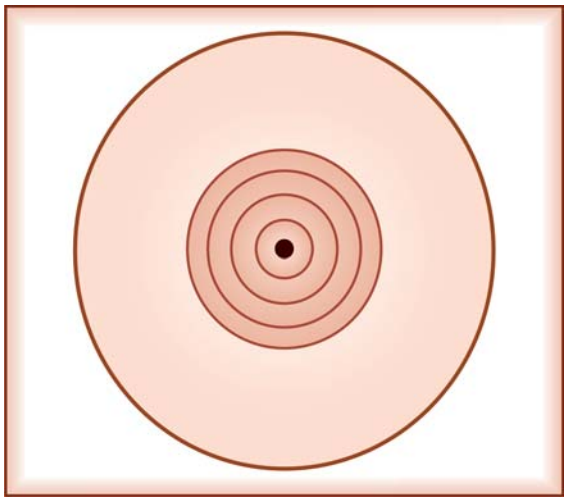


Fig. 9.9: Placido's disk reflex on the normal corneal surface

Corneal Topography

Corneal topography is a computerized videokeratography in which image of a Placido disk on the anterior surface of the cornea is captured by a

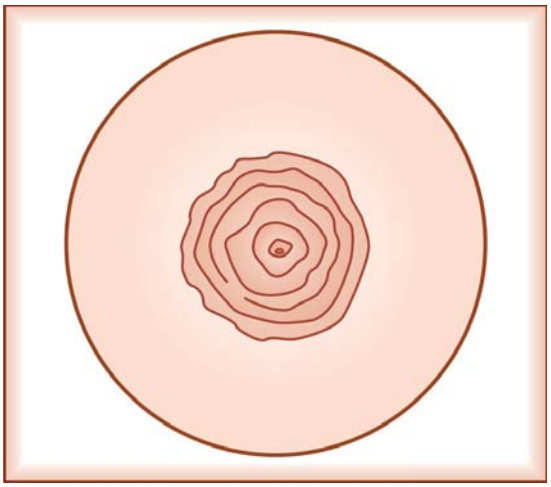


Fig. 9.10: Placido's disk reflex on irregular corneal surface

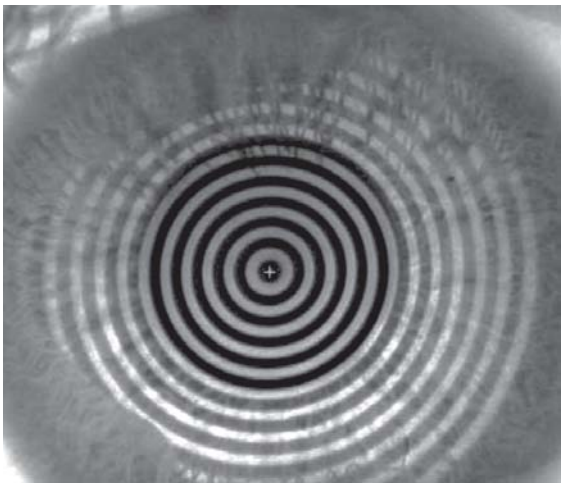


Fig. 9.11: Topography of a normal cornea

video camera and analyzed by a computer software and presented in the form of colored maps. Different types of colors are used to indicate different power curvatures; green represents near normal, higher than normal is indicated by red while blue-green represents lower power. Color-coded maps provide a rapid visual method for clinical diagnosis but fail to provide numerical values necessary for the management. In Placido's disk method (Fig. 9.11) changes in the curvature of the cornea are quantified by assigning a diopteric value to the curved surface between adjacent rings. The topography of the normal cornea may show a round, oval (Fig. 9.12), symmetric or asymmetric bow-tie pattern. The central cornea is more accurately mapped than the peripheral.

The corneal topography can reinforce the data obtained from patient's refraction, keratometry and slit-lamp examination and it is very useful in the detection of corneal pathologies such as early keratoconus, pellucid marginal corneal degeneration, keratoglobus and corneal dystrophies. It helps in contact lens fitting and calculation of intraocular lens power for implantation. Corneal topography guides the surgeon to plan for refractive surgery.

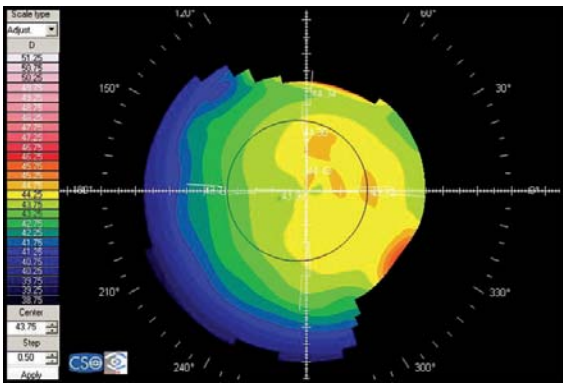


Fig. 9.12: Topography of a normal cornea showing oval pattern

Staining of Cornea with Vital Dyes

An abrasion in the cornea occurs more often than recognized. It causes discomfort, photophobia and watering. It may be easily overlooked if careful examination is not carried out. An inspection may not reveal an appreciable change in the bright corneal reflex. But corneal staining with vital dyes like fluorescein or rose bengal will confirm the abrasion. The application of fluorescein (2%) delineates the area of denuded epithelium, which takes brilliant green color. Rose bengal (1%) stains the devitalized cells as red or pinkish-red.

Corneal Opacities

The opacification of the cornea is a sequel to its inflammation. The opacites may vary from a mere nebula to a gross leukoma. If the corneal scar is thin it is called *nebula* (Fig. 9.13); it requires careful examination with a magnifying loupe or a slit-lamp. A nebula covering the pupillary area disturbs the vision more than a localized dense leukoma, since it refracts the light irregularly, whereas the leukoma stops all the light rays.

The destruction of the anterior layers of corneal stroma produces a dense opacity called *macula* (Fig. 9.14). If the opacity is very dense and white, it is termed as *leukoma* (Fig. 9.15).

Sometimes, iris is adherent to the back of a leukomatous opacity associated with irregular depth of the anterior chamber. The condition is called *leukoma adherence* which almost always occurs following perforation of the peripheral cornea. The situation and the extent of the corneal opacity in relation to the pupil and the limbus must be ascertained. Opacities situated away from the pupil seldom cause serious visual impairment.Vascularization of the opacity suggests an active lesion.

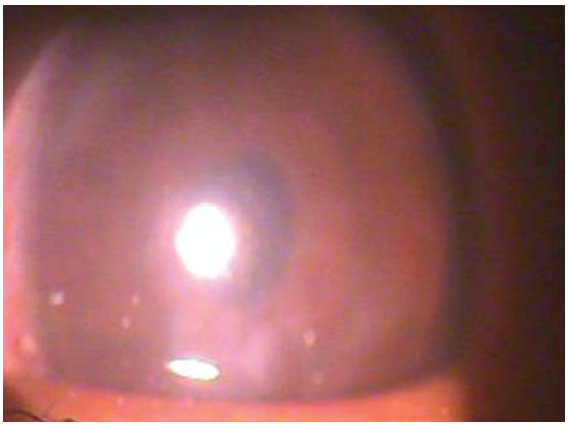


Fig. 9.13: Nebular corneal opacity

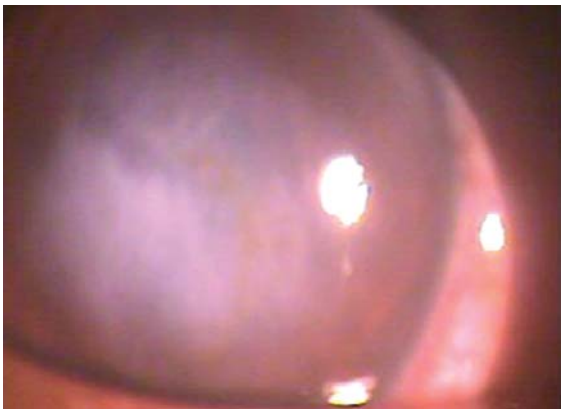


Fig. 9.14: Macular corneal opacity

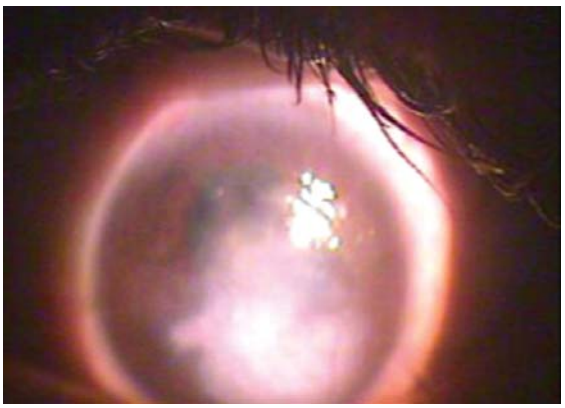


Fig. 9.15: Leukomatous corneal opacity

Vascularization of Cornea

The vascularization of cornea may be superficial or deep or sometimes mixed (Fig. 9.16).

The superficial and deep vascularization of cornea can be distinguished on the following points.

1. The continuity of superficial vessels can be traced over the limbus into the conjunctiva. However, deep vessels stop abruptly at the limbus.
2. The superficial vessels have a bright red color, while the deeper ones are dull red or grayish-red.

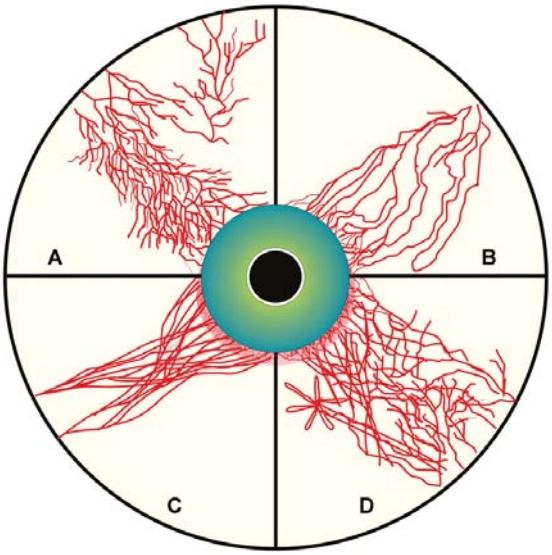


Fig. 9.16: Corneal vascularization: (A) Superficial type, (B) Terminal loop type, (C) Brush type, and (D) Mixed and umbel type

Fig. 9.17: Esthesiometer

3. The superficial vessels lie underneath the corneal epithelium and cause an unevenness of the surface, whereas deep vessels are buried in the corneal stroma and do not change the corneal surface.
4. The superficial vessels branch in an arborescent or dendritic pattern, but deep vessels run parallel to each other in a radial manner.

Esthesiometry

The cornea is richly supplied by nerves and is a very sensitive structure. The sensitivity can be tested by touching it with a wisp of cotton-wool and looking for blink reflex as a response. The cotton-wool should be brought from the side so as to avoid blinking in response to the menace reflex (reflex blinking due to an unexpected object coming all of a sudden in the near field of vision).

A more qualitative way of measuring corneal sensation is to use a Cochet-Bonnet esthesiometer in which a thin nylon monofilament is used for the stimulus (Fig. 9.17). Normally, the cornea is most sensitive in the center. The corneal sensation is often diminished in herpes, leprosy, neuroparalytic keratitis, absolute glaucoma and cerebello-pontine angle tumor.

Slit-lamp Examination of Cornea

Many corneal lesions like micropannus of trachoma, avascular superficial keratitis, subepithelial punctate keratitis and keratic precipitates (KPs) can best be studied with the help of a slit-lamp.

The slit-lamp (Fig. 9.18) consists of a binocular microscope with a brilliant light source which can be brought to a focus as a slit. The optical section of a normal cornea forms a *parallelepiped*, the brighter area corresponds to the surface and the darker to the deeper section of the cornea.

The micropannus is an extension of secondary corneal loops of vessels between the epithelium and Bowman's membrane alongwith distal infiltration. Avascular keratitis and dendritic lesions of herpes and Thygeson's superficial punctate keratitis presents a characteristic appearance if visualized on slit-lamp after staining the

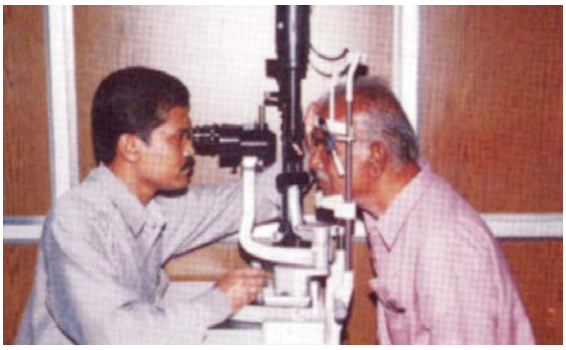


Fig. 9.18: Slit-lamp examination (*Courtesy*: Drs Anup Chirayath and Jyoti Anup, Aravind Eye Hospital, Tirunelveli)

cornea with fluorescein. The distinction between the superficial and the deep vascularization of cornea is easily made by slit-lamp biomicroscopy.

The keratic precipitates are accumulation of cells which adhere to the corneal endothelium and are diagnostic of anterior uveitis (iridocyclitis). The fresh KPs appear as small, round shining dots (composed of either lymphocytes or macrophages) adherent to the edematous corneal endothelium (Fig. 9.19). Many macrophagic keratic precipitates coalesce, and tend to form a triangular area on the inferior corneal endothelium (*Arlt's triangle*) resembling a mutton-fat (*mutton-fat KPs*). The presence of brown, shrunken KPs with crenated edges suggests old iridocyclitis.

Specular Microscopy

The corneal endothelium *in vivo* can be examined by a specular microscope. The instrument enables to take clear photographs and count the endothelial cells. The normal cell count is more than 3500 cells/mm^2 in children and decreases to 2000 cells/mm^2 in old age. The average cell count is 2400 cells per square millimeter (Fig. 9.20). Cornea with less than 1000 cells/mm^2 may not tolerate an intraocular surgery. Most of the endothelial cells have hexagonal shape. Variability in the shape of cells is called *pleomorphism*. The presence of more than 50% non-hexagonal cells is a contraindication for intraocular surgery.

Corneal Pachymetry

Corneal pachymetry measures the thickness of the cornea. Thickness of cornea varies from center to periphery. The central part of a normal cornea is between 520 µm and 560 µm thick. The peripheral zone has thickness between 630 µm and 670 µm. The superior cornea, in all the zones, is thicker than the inferior. Central corneal thickness (CCT) can be measured by ultrasonic pachymeter, laser interferometer or by optical coherence tomography. CCT is increased in acute or chronic corneal edema caused by traumatic, inflammatory and dystrophic conditions. Corneal thickness can

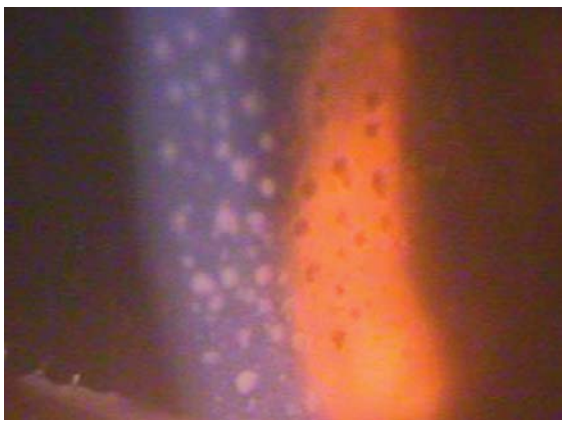


Fig. 9.19: Keratic precipitates

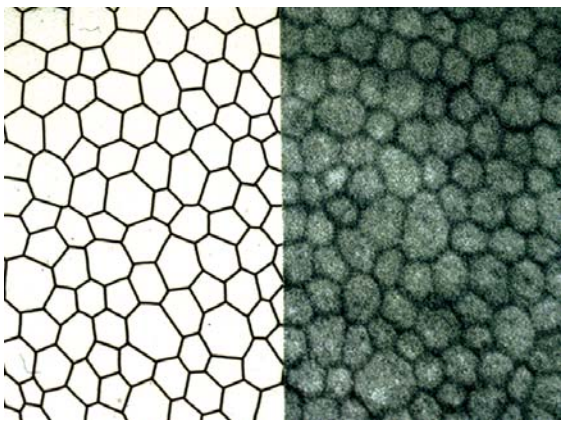


Fig. 9.20: Corneal endothelium (*Courtesy:* Mr S Kanagami, Tokyo)

alter the measurement of intraocular pressure (IOP) by applanation tonometer. Patients with increased central corneal thickness (> 600 µm) record artificially high IOP while those with decreased central corneal thickness (< 500 µm) record low IOP.

Examination of the Sclera

The sclera cannot be visualized directly as it is covered by episclera and conjunctiva. A raised, localized, bluish or purple, congested nodule slightly away from the limbus suggests *episcleritis*, while a diffuse, dusky patch with ciliary congestion points to the involvement of the sclera called *scleritis*. The latter may cause a tongue-shaped opacity in the deeper layers of the cornea at the periphery, *sclerosing keratitis*. Occasionally, *blue discoloration of the sclera* is found as an isolated anomaly or may be associated with osteitis deformans. Pigmentation of the sclera is also seen in *nevus of Ota* and *melanosis bulbi*. A localized ectasia of the sclera associated with herniation of the ciliary body, known as *ciliary staphyloma*, is often seen following scleritis or trauma. A ring ciliary staphyloma is not unusual in longstanding cases of buphthalmos or secondary glaucoma in children.

Examination of the Anterior Chamber

The anterior chamber is nearly 2.5 mm deep in the center. The depth is estimated by the position of the iris and is easily determined by oblique illumination of the anterior segment of the eye. The inclined beam of light illuminates the nearby iris surface but the transpupillary surface of the iris remains dark in shallow chamber. When the chamber depth is normal or deep, both nasal and temporal surfaces of the iris are equally illuminated.

The anterior chamber is shallow in extremes of ages, angle-closure glaucoma and high hypermetropia. A deep anterior chamber is found in high myopia, buphthalmos, aphakia, posterior dislocation of lens and keratoglobus. The chamber is frequently unequal in depth in iridiocyclitis (shallow at the periphery and deep in the center), anterior synechia and anterior subluxation of the lens.

Fig. 9.21: Aqueous flare (fine particles) in the anterior chamber

The anterior chamber is filled with transparent aqueous humor. In acute iridocyclitis, the aqueous contains a number of inflammatory cells and protein and, therefore, becomes turbid. The presence of protein particles in the aqueous produces an *aqueous flare* which can be demonstrated by a narrow beam of light of slit-lamp (Fig. 9.21). Sometimes, in cases of corneal ulcer and/or acute iridocyclitis there occurs frank pus in the anterior chamber (*hypopyon*). The collection of blood in the anterior chamber (following ocular trauma, surgery, herpes zoster or gonococcal iridocyclitis) is known as *hyphema*. Occasionally, tumor cells from retinoblastoma or malignant melanoma may migrate into the anterior chamber and produce *pseudohypopyon*.

The Angle of the Anterior Chamber

The angle of the anterior chamber can be examined with the help of a gonioscope and slit-lamp (Fig. 9.22). Goldmann's gonioscope (Fig. 9.23) is a special type of contact lens fitted with mirrors in which the image of the recess of the angle is reflected. While examining the angle with a

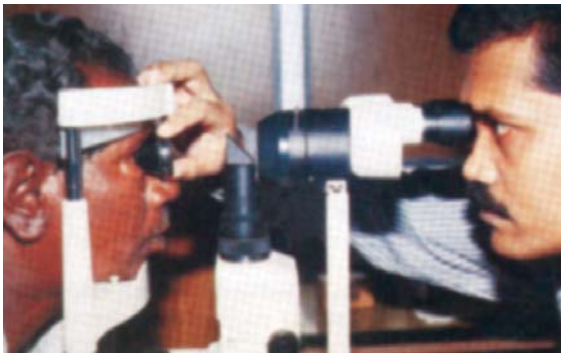


Fig. 9.22: Gonioscopy (*Courtesy*: Drs Anup Chirayath and Jyoti Anup, Aravind Eye Hospital, Tirunelveli)

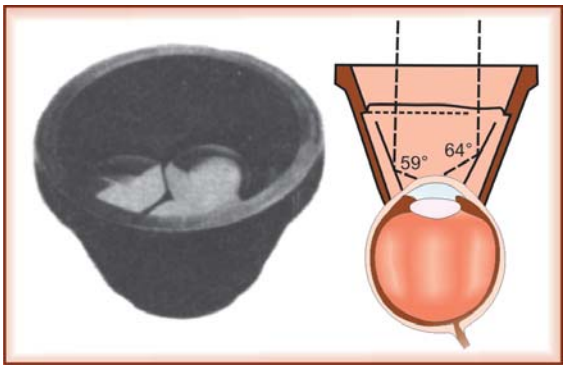
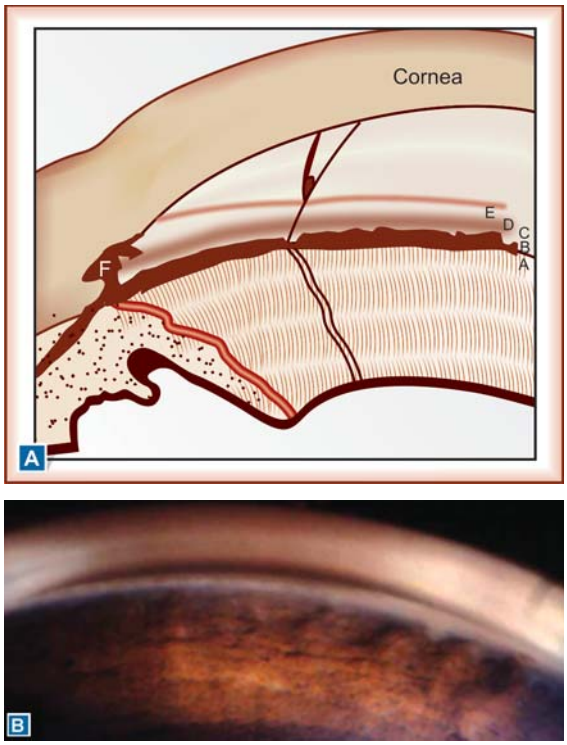


Fig. 9.23: Goldmann's 3-mirror gonioscope



Figs 9.24A and B: (A) Anatomical landmarks of the angle of the anterior chamber: A: iris root, B: ciliary body, C: scleral spur, D: trabecular meshwork, E: Schwalbe's line, F: Schlemm's canal, (B) Gonioscopic view of the angle

narrow slit of light, a 'V' of light is seen. One leg of the 'V' outlines the corneal surface of the angle and the other the iris surface. The important landmarks of the angle from behind forward are the root of the iris, the anteromedial surface of the ciliary body, the scleral spur, the trabeculum with the canal of Schlemm, the Schwalbe's line and the posterior surface of cornea (Figs 9.24A and B). Depending on the visibility of these structures, the width of the angle of the anterior chamber can be graded as suggested by Shaffer (Fig. 9.25).

The configuration of the angle of the anterior chamber provides a basis for classifying glaucoma into two main categories—*open-angle* and *angle-closure glaucoma*. Gonioscopy helps in localizing a foreign body, abnormal blood vessel or tumor in the angle. It also demonstrates the presence of peripheral anterior synechiae and thus helps in planning the surgery for glaucoma.

van Herick's Slit-lamp Grading

The slit-lamp assessment of the depth of the peripheral anterior chamber (AC) and its comparison with the thickness of the overlying cornea can provide a fair estimate of the width of the angle of the anterior chamber even in the absence of gonioscopic examination.

Grading

Grade 0 represents an iris contact with the endothelium of the cornea; closed-angle.

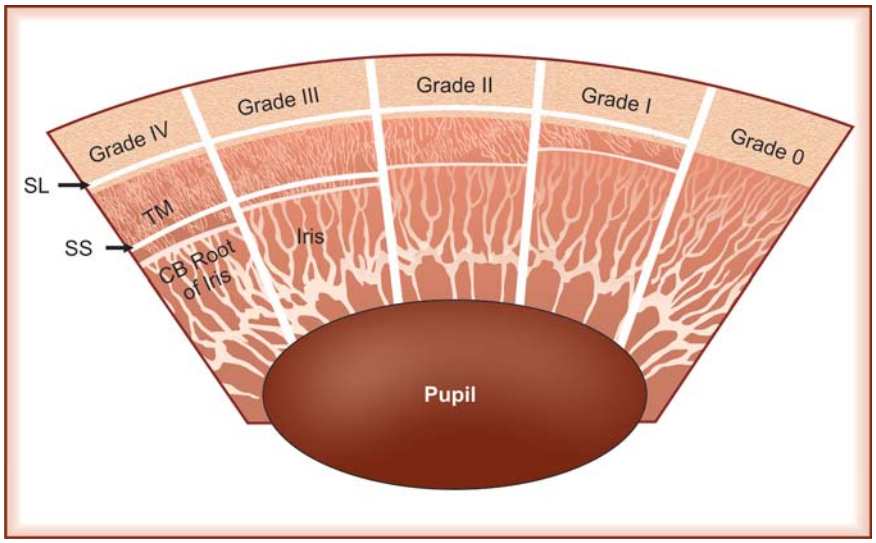


Fig. 9.25: Shaffer's grading of the angle of the anterior chamber—SL: Schwalbe's line, TM: Trabecular meshwork, SS: Scleral spur, CB: Ciliary body

Grade 1 represents the depth of the peripheral AC less than 1/4th thickness of the cornea; narrow-angle.

Grade 2 represents the depth of the peripheral AC between 1/4th and 1/2 the corneal thickness; open-angle.

Grade 3 represents the depth of the peripheral AC more than 1/2 the corneal thickness; wide open-angle.

Examination of the Iris

The color of the iris varies from individual to individual, it is light blue or green in Caucasians and dark brown in Orientals. The two irides or a sector of the same iris may be of different colors—*heterochromia*. Generally, the surface of the iris is shining and transparent revealing the collarette and crypts, but in iridocyclitis the iris appears dull and muddy obscuring the normal pattern due to inflammatory exudates. Sometimes, tags of iris tissue may remain adherent to the collarette (*persistent pupillary membrane*).

Gray or white patches on the iris are tell-tale signs of chronic iridocyclitis or acute congestive glaucoma. A gap or hole in the upper sector of the iris suggests surgical coloboma, while its presence in the lower sector is often due to a defective development, congenital coloboma. Melanoma, tuberculoma, gumma and sarcoidosis may manifest as raised nodules on the iris surface. Abnormal vascularization of the iris is often seen in diabetes, occlusion of the central retinal vein and melanoma of the iris.

The plane of the iris is found to be disturbed in several pathological entities. A forward bowing of the iris (*iris bombé*) is a sign of early angle-closure glaucoma. Adhesion of the iris with the cornea (*anterior synechia*) is a common sequel to perforation of the corneal ulcer. *Posterior synechia* (adhesion of the iris with the lens) is frequently seen in iridocyclitis. Normally, iris rests on the anterior surface of the lens, but this support is lost in aphakia resulting in tremulousness of the iris (*iridodonesis*).

Examination of the Pupil

The pupil is a circular aperture of about 4 mm diameter nearly in the center of the iris,placed slightly nasally (Fig. 9.26). The pupillary size remains in a continuous state of flux adjusting to the change in ambient illumination and fixation distance. It tends to be smaller in infants and elderly persons than in young adults, and smaller in brown eyes than in blue eyes. Constriction of the pupil is known as *miosis*, while dilatation as *mydriasis*. Rarely, there can be more than one pupillary aperture called *polycoria*. Occasionally, the location of pupil may be eccentric (*corectopia*).

Pupillary Light Reactions

Normal pupil reacts to light directly or consensually as well as to convergence and accommodation.

Direct light reaction is elicited by keeping the patient in a dark room and asking him to fix gaze at a distant object to prevent activation of the near reflex. A narrow beam of light is thrown on the eye while watching the pupil. The same procedure is repeated in the other eye. A normal pupil reacts briskly to the light and its constriction remains sustained unless the light source is removed. An ill-sustained pupillary reaction (*Marcus-Gunn pupil*) is found in retrobulbar neuritis owing to afferent conduction defect.

Consensual light reaction is demonstrated by exposing only one eye to the light (blocking the light from the other eye by keeping the palm at the level of nose) and watching the pupillary reaction in other eye. Normally, the pupil reacts briskly. The same procedure is repeated in the other eye.

The *swinging flashlight test* is performed by asking the patient to sit in a room with diffuse background illumination. A bright light from an indirect ophthalmoscope is directed briskly and rhythmically from eye-to-eye several times and differences in pupillary reaction, if any, are noted. Normally both pupils constrict equally. However, in the presence of a *relative afferent pupillary defect* (RAPD), the affected pupil shows reduced amplitude of constriction and accelerated dilatation (recovery) as compared with the contralateral eye (control). RAPD suggests the presence of a unilateral or asymmetric optic nerve disease (Fig. 9.27).

Near reaction is a synkinesis consisting of convergence, accommodation and pupillary constriction (miosis). The reaction to convergence and accommodation is determined by asking the patient to focus on a far point and then telling him/her to look at a pencil brought near to the eye suddenly and held 15 cm away—normally the pupils constrict while the eyes converge. If the pupillary

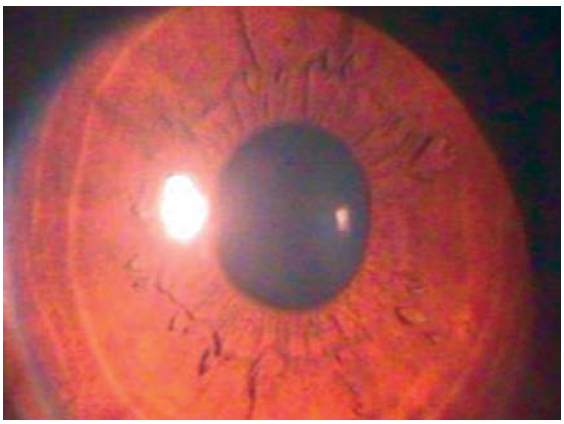


Fig. 9.26: Normal pupil

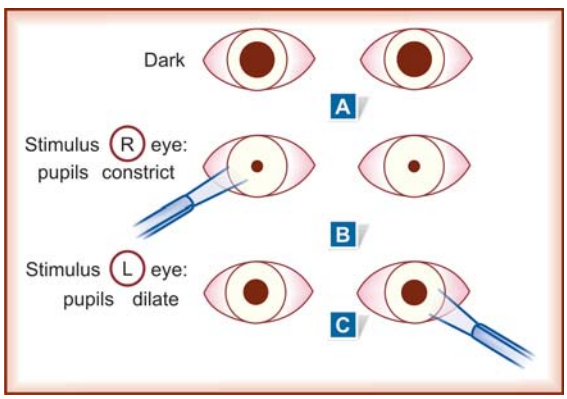


Fig. 9.27: Swinging flash light test in a patient with left optic nerve lesion

response to light differs significantly than that to near stimulus, the condition is called *light-near dissociation*. It is found in the lesions of anterior visual pathway, diabetes mellitus, and pretectal lesions.

Abnormal pupillary reactions are encountered in the lesions of the visual pathway and the common ones are described here.

Hemianopic pupillary reaction (Wernicke's) can be elicited by a narrow beam of slit-lamp, the pupil reacts briskly when the intact half of the retina is illuminated but does not react when the other half is illuminated. The syphilitic lesion of the tectum affecting the pupillary pathway often results in *Argyll-Robertson pupil* wherein the pupils are small and the light reaction is impaired, but the reaction to convergence and accommodation is retained. Sometimes, a unilateral dilated but tonic pupil (*Adie's pupil*) of unknown etiology is found in young women associated with loss of knee-jerk. Apparently a tonic pupil does not react to light and convergence, but careful examination reveals a very sluggish reaction with long latent period. Adie's pupil can be differentiated from Argyll-Robertson pupil as the former dilates well with atropine while the latter does not.

Small constricted pupils (miosis) are found in persons using topical miotics or systemic morphine. Other causes of small constricted pupils are irritation of third nerve by pontine hemorrhage and sympathetic paralysis. A unilateral miosis due to sympathetic paralysis is accompanied with narrowing of palpebral fissure, slight enophthalmos and unilateral absence of sweating. This condition is called *Horner's syndrome*.

Dilatation of pupils (mydriasis) occurs after instillation of mydriatic or cycloplegic drugs and the patient often has difficulty in doing near work. The common causes of pupillary dilatation are—acute congestive glaucoma (large vertically oval pupil), absolute glaucoma, optic atrophy, third nerve palsy (ophthalmoplegia interna) following meningitis, encephalitis, lead poisoning or orbital trauma. Sometimes, unilateral dilatation of pupil may occur from irritation of the cervical sympathetics by enlarged cervical glands in the neck, apical pleurisy, cervical rib, meningitis and acute anterior poliomyelitis. Many of these cases subsequently develop pupillary constriction from sympathetic paralysis.

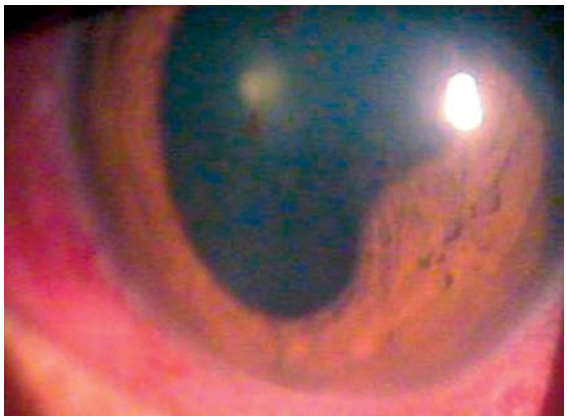


Fig. 9.28: Festooned pupil

A small, irregular, sluggishly reacting or immobile pupil associated with muddiness of the iris is the hallmark of *iritis*. Irregularity and immobility of the pupil occur due to posterior synechiae, and instillation of a mydriatic results in a festooned or pear-shaped incompletely dilated pupil (Fig. 9.28). It is not rare to find ectropion of pigments over the pupillary border after anterior uveitis. Rarely, the pupillary aperture is completely occluded by exudates—*occlusio pupillae*.

When the light is thrown upon the pupil of a young person's eye, it appears bluish-black. The pupil of an adult looks gray owing to the scattering of light from the lens surface. An old person's pupil may look brown or white due to cataract formation. A white pupillary reflex in infants and young adults must arouse the suspicion of retinoblastoma and pseudoglioma, respectively.

Examination of the Lens

The lens of the eye is a transparent structure and, as such, cannot be thoroughly examined without

the help of a slit-lamp. The pupil must be fully dilated before the examination of the lens. On focal illumination, the lens of youngsters appears almost clear or gives a faint bluish hue. However, in old persons, it imparts gray-white or yellow reflex. Mere presence of lenticular haze by oblique illumination does not warrant the diagnosis of cataract as the refractive index of the lens increases with the age and causes marked scattering of light contributing to the haze. An ophthalmoscopic examination often gives a clear red reflex. Therefore, the findings must be confirmed either by ophthalmoscopic examination or by distant direct examination by a plane mirror.

The slit-lamp examination provides an optical section of the lens which shows from within outward—embryonic nucleus, fetal nucleus, infantile nucleus, adult nucleus, cortex and capsule (Fig. 9.29). An anterior 'Y'-shaped and a posterior inverted 'Y'-shaped sutures are also seen.

Any opacity in the lens is called *cataract* which may be either *developmental* or *acquired*. The developmental cataract affects a particular zone of the lens and may or may not cause impairment of vision. Among the acquired cataracts, senile cataract is the commonest which may manifest either in a cortical or a nuclear form. *Cortical cataract* (Fig. 9.30) starts as triangular spokes of opacities with their apices towards the center of pupil. They coalesce and form a white, more or less, total opacity also known as *soft cataract*. An accentuation of the process of lenticular sclerosis results in *nuclear cataract* (Fig. 9.31) which often presents a brown or black central reflex. The lenticular opacities appear black against a red fundus reflex when viewed by a plane mirror or an ophthalmoscope. Occasionally, the lens may be transparent but a white reflex in the pupil (*leukocoria*) is found due to retinoblastoma, total retinal detachment, tuberculoma of the choroid, Coats' disease, persistent hyperplastic primary vitreous and retrolental exudative membrane.

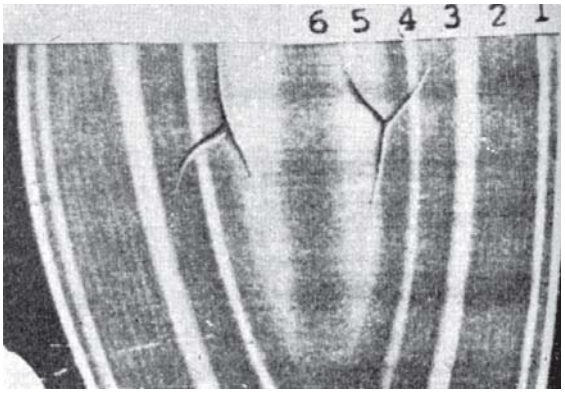


Fig. 9.29: Optical section of the adult lens 1. Capsule, 2. Cortex, 3. Adult nucleus, 4. Infantile nucleus, 5. Fetal nucleus, 6. Embryonic nucleus

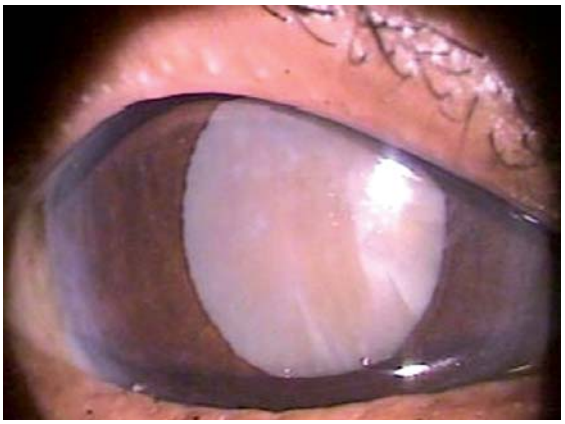


Fig. 9.30: Cortical cataract

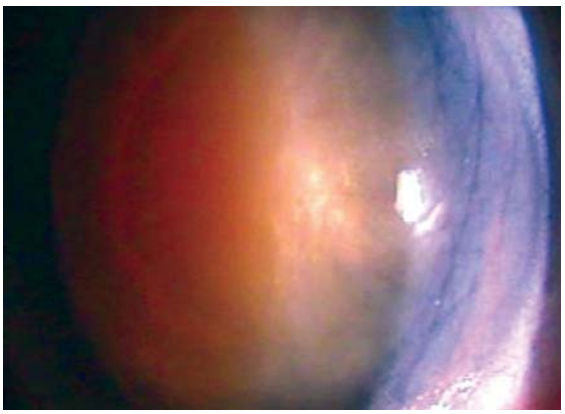


Fig. 9.31: Nuclear cataract

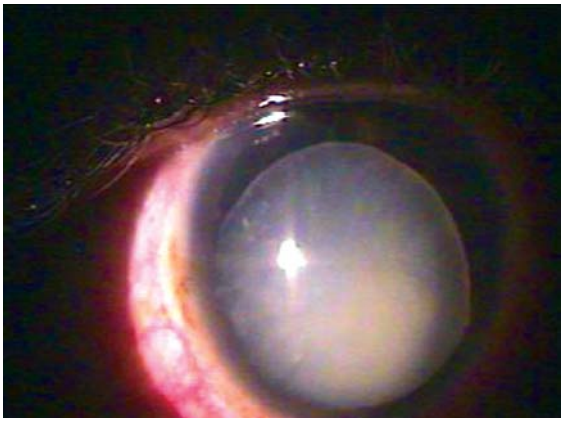


Fig. 9.32: Subluxated lens

The lens is kept in its position by the suspensory ligaments. If the ligaments are weak or improperly developed, the lens gets subluxated (Fig. 9.32) as seen in Marfan's syndrome and homocystinuria. In Weill-Marchesani's syndrome the lens is small and spherical (*microspherophakia*) and, hence, wanders in the pupillary area. The subluxated lens can be diagnosed by segmental tremulousness of the iris, presence of the edge of the lens in the pupillary area and demonstration of phakic and aphakic areas by retinoscopy or ophthalmoscopy. The lens may be dislocated in the vitreous, anterior chamber or subconjunctival space following trauma. A posterior dislocation or subluxation can also occur spontaneously in hypermature cataract. Couching used to be a cause of posterior dislocation of the lens in our country. Deep anterior chamber, tremulousness of the iris and floating lens in the vitreous are the characteristic signs of the posterior dislocation of the lens.

When a strong beam of light falls on the eye, four images (*Purkinje's images*) are formed on the four reflecting surfaces—anterior surface of the cornea, posterior surface of the cornea, anterior surface of the lens and posterior surface of the lens. As the first three surfaces are convex, therefore, if the light source moves, the images also move in the same direction; the fourth image moves in the opposite direction since the fourth surface is concave. Usually, the first and the fourth images are clearly visible on pupillary dilatation. However, the second and third images can only be recognized in a dark room with the help of a brilliant light. The presence of the fourth Purkinje's image indicates that the lens is clear. In cataract the fourth image is absent but third can be demonstrated. Both third and fourth Purkinje's images are absent in aphakia.

Ocular Tension

Ocular tension is the record of the intraocular pressure (IOP). The latter can correctly be measured by manometry. Since manometric measurements are not practical in human beings, indentation or flattening of a limited area of the cornea by a given weight is measured. The technique is known as *tonometry*. The tension can be assessed roughly by digital tonometry (Fig. 9.33). The patient is asked to look down towards his feet and tips of the index fingers of the examiner are placed side-by-side on the upper lid just above the upper border of the tarsal plate. One finger is kept stationary while the other presses to indent the globe and an impression of fluctuation is felt by the stationary finger. A normal fluctuation can only be appreciated by practice. If the tension is high, the fluctuation is feeble or

Fig. 9.33: Digital tonometry

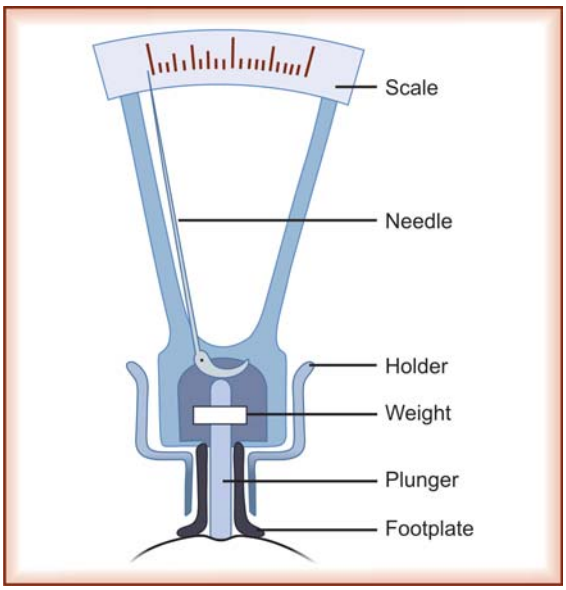


Fig. 9.34: Schiotz's tonometer

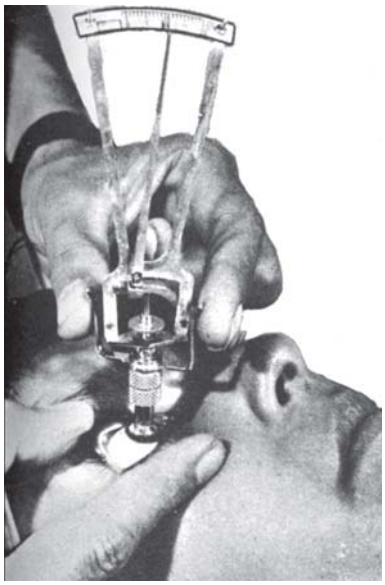


Fig. 9.35: Schiotz's tonometry

absent. An eye having low tension gives a feeling of a water-bag.

An accurate determination of ocular tension is made by means of an instrument called *tonometer*. Different types of tonometers are in use. For indentation tonometry, Schiotz's tonometer (Fig. 9.34) is most commonly used. The tonometer measures the amount of indentation of the cornea produced by the weight of its plunger or any additional weights on it, which is indicated by the deflection of a pointer on a graduated scale. The instrument is calibrated so that the equivalent of readings in millimeter of mercury (mm Hg) can be read from a chart. Ocular tension measured by the tonometer is inaccurate because of wide individual variation in the rigidity of the sclera. The error can be minimized by differential tonometry (using different weights 5.5, 7.5 and 10 g) and obtaining a correct IOP from Friedenwald's nomogram.

Technique of Recording Ocular Tension

Schiotz's tonometry is done in lying position and an anesthetic solution (lignocaine 4% or proparacaine 0.5%) is instilled into each eye. The patient is asked to see a spot on the ceiling with both the eyes. Both upper and lower eyelids are separated with the fingers and a Schiotz's tonometer is placed vertically on the corneal surface of each eye so that it rests by its own weight (Fig. 9.35). The scale reading is taken from the tonometer. Further readings are obtained by putting additional weights to make the final weight of the plunger 7.5 and 10 gm. The intraocular pressure is then determined by the nomogram. The ocular tension by Schiotz's tonometer varies between 12 and 20 mm Hg in a normal individual. A rise of more than 21 mm Hg should arouse a suspicion of glaucoma. Besides scleral rigidity, a poorly calibrated tonometer, improper positioning of the tonometer on the cornea and an uncooperative patient are the common sources of error. Therefore, Schiotz's tonometry is considered obsolete and applanation tonometry is preferred.

Applanation tonometry is widely used in ophthalmic practice since it eliminates the factor of ocular rigidity and records the tension more accurately than the Schiotz's. It records the force required to flatten a small constant area on the central cornea

Fig. 9.36: Applanation tonometry (*Courtesy:* Drs R Ramakrishanan and Datta Ravi, Aravind Eye Hospital, Tirunelveli)

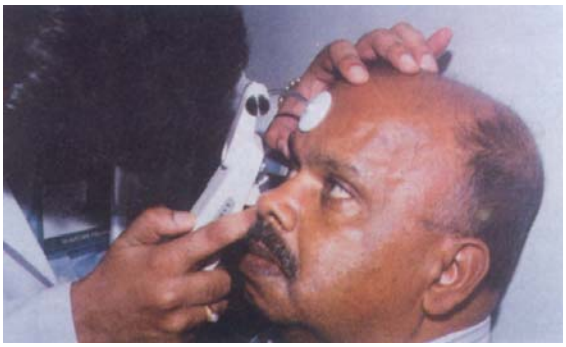


Fig. 9.37: Hand-held Perkin's applanation tonometer (*Courtesy:* Drs R Ramakrishanan and Datta Ravi, Aravind Eye Hospital, Tirunelveli)

which is directly proportional to the intraocular pressure. The applanation tonometer is generally used with a slit-lamp (Fig. 9.36). The ocular tension recorded by applanation tonometer ranges between 14 and 17 mm Hg, with an average of 15 mm Hg. A hand-held Perkin's applanation tonometer can also be used (Fig. 9.37).

A number of other tonometers like air-puff tonometer, tono-pen and pneumotonometer are available for recording the IOP.

Air-Puff tonometer (Noncontact tonometer) does not require topical anesthesia. The noncontact tonometer deforms the corneal apex by a pulsed jet of air. The time required to flatten the cornea relates directly to the level of IOP.

Tono-Pen has a software that measures the IOP accurately. It is preferred in the patients with scarred cornea. To minimize the error, at least three IOP measurements should be taken.

Pneumotonometry is a useful device to measure IOP in eyes with irregular corneal surface or scarred cornea. The sensor measures the air pressure by indenting the cornea by a flow of gas against a flexible diaphragm. It provides a record of IOP on a paper strip.

Transillumination

Transillumination is an excellent method for localizing the tumors of ciliary body or anterior choroid. It is usually of two types—*trans-conjunctivoscleral* and *transcorneal*. In the former, an intense beam of light is thrown from the fornicial conjunctiva and sclera, and pupil is visualized which normally appears red. However, if there lies a solid mass in the ciliary region, the pupil remains dark in the corresponding sector. The method can differentiate between a cyst and a tumor. The transillumination test can also be performed for the posterior segment lesions by inserting a fiberoptic transilluminator after opening the Tenon's capsule.

When the cornea and the lens are opaque, transcorneal transillumination can be of help to localize lesions of the ciliary body. The test is carried out in a dark room. The pupil is dilated and the cornea is anesthetized. The cornea is covered with a rubber cap attached to the transilluminator. The entire eyeball illuminates except for the corona ciliaris and vitreous base. A tumor or foreign body in the ciliary body or choroid will appear dark.

Ophthalmoscopic Examination or Ophthalmoscopy

The routine ophthalmoscopic examinations performed in a dark room are listed below.

1. Preliminary examination with a mirror at 22 cm distance (*distant direct ophthalmoscopy*)
2. Ophthalmoscopic examination by indirect method (*indirect ophthalmoscopy*)
3. Ophthalmoscopic examination by direct method (*direct ophthalmoscopy*).

Generally, examination with a mirror at one meter distance gives information about the nature of the refractive status of the eye under examination, known as *retinoscopy*, and is already been covered in the Chapter on *Determination of the Refraction*.

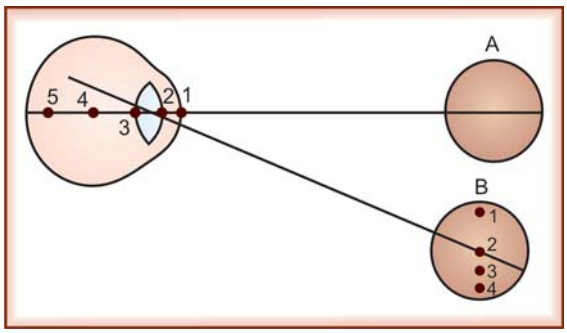


Fig. 9.39: Parallactic displacement

Distant Direct Ophthalmoscopy

Preliminary examination with a mirror at 22 cm distance is a useful method (Fig. 9.38) which provides the information about opacities in refracting media and the status of the retina. It also confirms the findings of the external examination of the eye. It is carried out in a dark room. When the light is thrown with a plane mirror, the fundus reflex is normally seen as a uniform red glow. If there are opacities in the media they appear black against the red background. The opacities may be stationary or floating. The nature of the opacities can be determined by asking the patient to move his eyes; opacities in the vitreous and aqueous will continue to move even after the eye is brought to rest. The exact position of the opacity can be determined by observing its parallactic displacement (Fig. 9.39). By this method the opacities in the pupillary plane will appear stationary, those in front of that plane (cornea and anterior chamber) will move in the same direction, and those behind the plane (posterior surface of the lens and vitreous) will appear to move in opposite direction.

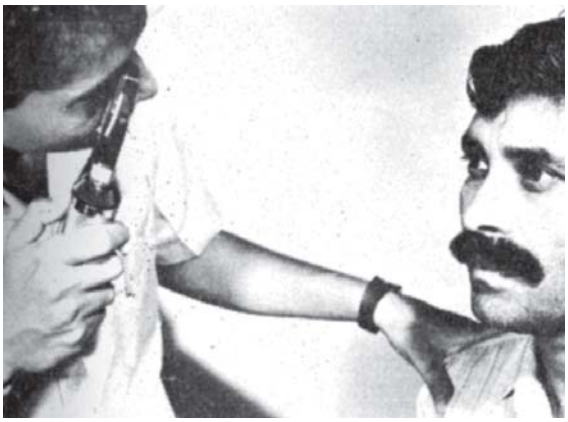


Fig. 9.38: Distant direct ophthalmoscopy

The plane mirror examination clearly distinguishes between an iris hole and a mole and locates the edge of a dislocated lens or congenital coloboma of the lens. The edge of the lens appears as a black crescent in the pupillary area due to total reflection of light within the lens. The presence of a whitish or grayish uneven surface with black retinal vessels suggests a detached retina or a tumor arising from the retina. In place of the mirror, a self-illuminated ophthalmoscope can be used for the examination.

Indirect Ophthalmoscopy

The indirect ophthalmoscopic examination is an important procedure to examine the details of the fundus, particularly of the periphery (Fig. 9.40). The optical principle utilized in this method is to make the eye highly myopic by placing a strong convex lens (+20 D spherical) about 10 cm in front of the eye so that a real inverted aerial image of the fundus, magnified about three times, is formed

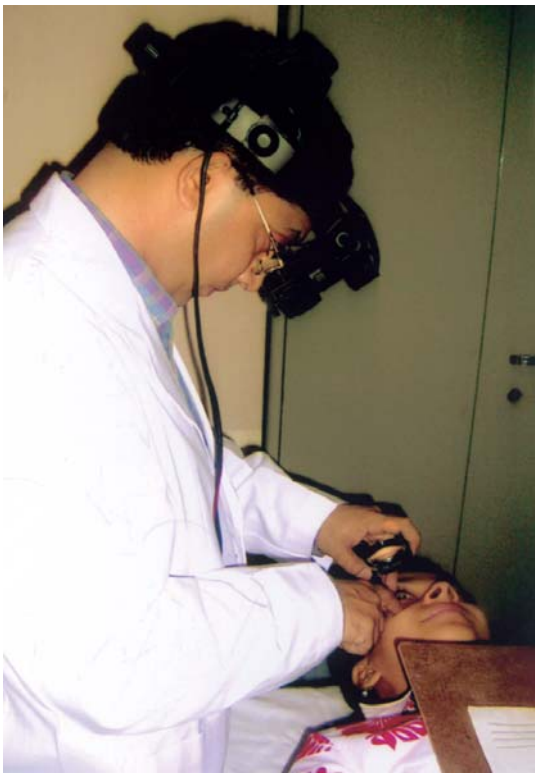


Fig. 9.40: Indirect ophthalmoscopy

between the observer and the convex lens. In case the lens is kept at a constant distance from the eye (at its own focal distance), the image will be formed at the focal distance of the lens in emmetropic eye, near the lens in myopic eye and farther from it in hypermetropic eye.

The examination requires practice and the examiner has to adjust the distance between the lens and the patient's eye by moving the lens forward or backward until a clear view of the fundus is obtained. The optic disk of right eye can easily be focused by asking the patient to look towards right and for the left optic disk he should fix the gaze on his left side. For the examination of the macula, the patient may be asked to look in the light, and the periphery of the fundus can be explored by asking the patient to look upwards, downwards and sidewards. Ora serrata can be visualized by scleral indentation. Corneal reflexes and reflections from the condensing lens often obscure the retinal image if the lens is kept at its focal length from the patient's eye. Therefore, a slight tilting and shifting of the lens will provide greater clarity to the observer.

Indirect ophthalmoscopy is superior to the direct ophthalmoscopy as it provides a large field of vision even through a semi-dilated pupil and a good view of fundus is not affected by opacities in the media or by the presence of high refractive errors (Table 9.2).

Table 9.2: Comparison of indirect and direct ophthalmoscopy

Features	*Indirect*	*Direct*
1. Image formation	True and inverted	Virtual and erect
2. Magnification	3 to 5 times	About 15 times
3. Illumination	Bright (Funduscopy possible despite opacities in media)	Not so bright (Funduscopy not possible if opacities in media)
4. Stereopsis	Present	Absent
5. Area of field in focus	Nearly 8 disk-diameters	Only 2 disk-diameters
6. Accessible fundus view	Up to ora serrata	Up to equator

Direct Ophthalmoscopy

Hermann von Helmholtz reinvented the ophthalmoscope (Fig. 9.41) in 1851. The instrument has made it possible to examine living vascular and neural tissues under magnification. It has a unit of strong light that is directed into the patient's eye by reflection from a small mirror. The examiner's eye receives back the reflected light from the fundus of the patient through an aperture in the ophthalmoscope, thus an erect virtual magnified image of the fundus is obtained.

It is customary to dilate the pupil with a mydriatic like phenylephrine-tropicamide combination. The examiner stands to the side of the patient's eye

Fig. 9.41: Direct ophthalmoscope

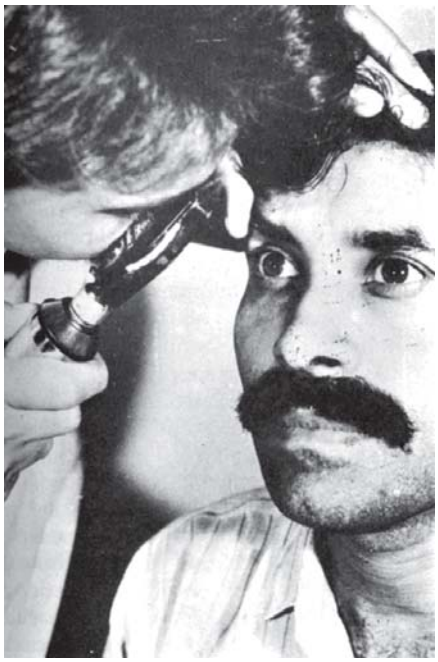


Fig. 9.42: Direct ophthalmoscopy

to be examined. For example, for the examination of the right eye he should be on the right side and use his right eye (Fig. 9.42). The aperture of the ophthalmoscope is held as close as possible to the observer's eye as well as to the patient's eye. If both examiner and patient are emmetropic, the reflecting rays will be parallel and easily focused on observer's retina. However, in order to get a clear image in ametropia, a correction of the refractive error must be made with the help of a set of lenses incorporated in the ophthalmoscope. The beginners may not be able to obtain a clear image even in an emmetropic eye since they find it difficult to relax their accommodation entirely at such a close distance. An addition of minus lenses may help them to see clearly.

For examination of the optic disk, the patient is advised to look straight ahead, while macula is observed by asking the patient to look at the light of the ophthalmoscope. Retinal blood vessels should be traced from the disk towards the periphery in all the sectors. The peripheral parts of the fundus (the areas adjacent to the equator) can be examined by asking the patient to look in different directions of gaze.

Direct ophthalmoscopy in emmetropic eye gives about 15 times magnified image of the fundus; the magnification is more in myopia and less in hypermetropia. Due to inequal magnification in two meridians the image is not clear in astigmatism.

Biomicroscopic Ophthalmoscopy

The Goldmann contact lens, the Hruby lens and high spherical +78 or +90 D hand-held lens can give detailed information about retinal disorders. Generally, slit-lamp biomicroscopy with +90 D lens is performed. It enables binocular viewing of the fundus and is useful in the diagnosis of macular lesions—edema, hole, cyst and subretinal neovascularization.

Fundus Oculi

The fundus of the eye appears bright red due to choroidal circulation. The details of fundus should be examined and recorded in a systematic way.

Optic Disk

The normal optic disk (optic nerve head) is about 1.5 mm in diameter, nearly circular with distinct margins (Fig. 9.43). Its color is pale-pink. There is a funnel-shaped depression in the central part of the disk known as *physiological cup*. The cup is usually shallow and covered by the meshes of lamina cribrosa. The central retinal vessels emerge from the middle of the cup. However, the extent of cupping and inclination of central vessels vary in different eyes. Surrounding the cup lies the neuroretinal rim of the disk which may show localized or generalized atrophy in glaucoma resulting in the enlargement of the cup. As the choroid and pigment epithelium fail to extend quite up to the margin of the disk, a narrow white ring of sclera is seen around the disk, the *scleral ring*. Occasionally, the black pigments from the pigment epithelium gather around the disk (crescent).

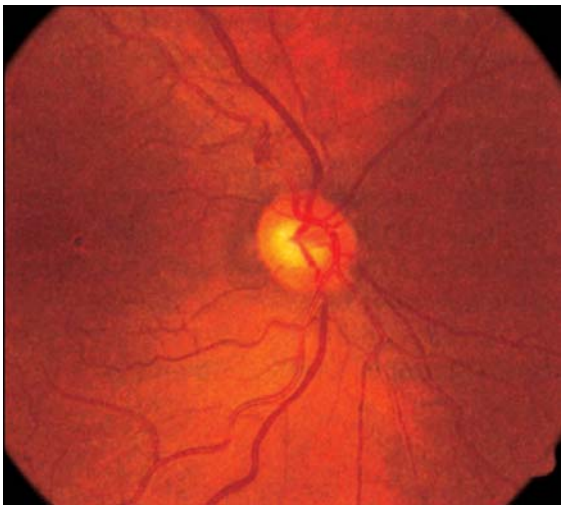


Fig. 9.43: Normal fundus

The optic disk may be implicated in the diseases of the eye as well as the central nervous system. The disk appears large in high myopia and small in high hypermetropia or aphakia. The disk becomes pale and atrophic in optic atrophy. In papilledema and papillitis there occurs blurring of the disk margin with obliteration of the cup, but in glaucoma and arteriosclerosis the cup enlarges. Pseudopapillitis is seen in high hypermetropia and astigmatism.

Macula

The macula is situated at the posterior pole about 2 disk diameters (3 mm) lateral to the temporal margin of the disk, slightly below the horizontal meridian. The pupil should be well-dilated before the commencement of examination of the macula. The macular area appears darker than the surrounding fundus. It is circular and has a diameter of approximately 5 mm. The center of the macula is known as *fovea centralis* which imparts a bright reflex (foveal reflex) due to reflection of light from the walls of the foveal pit. The fovea is an avascular area mainly supplied by the choriocapillaris. The blood supply of macular region is through small twigs from the superior and inferior temporal branches of the central retinal artery. These twigs run radially and terminate nearly 0.5 mm away from the fovea. Sometimes, a cilioretinal artery, originating in a hook-shaped manner within the temporal margin of the disk, runs towards the macula to supply it.

Diseases of the macula are common and they severely impair the central vision. Punched-out macular lesions are found in congenital toxoplasmosis. The foveal reflex is dull or lost in central retinochoroiditis or macular edema following trauma, eclipse burn and central serous retinopathy. Resolution of edema results in a granular appearance of the macula associated with loss of foveal reflex. A macular scar is found in healed central chorioretinitis. A macular star is seen in patients with uncontrolled hypertension or

papilledema. A small, circular retinal cyst at the macula may be confused either with a round hemorrhage or with a macular hole. Additional examination by slit-lamp biomicroscopy, fluorescein angiography and optical coherence tomography are required to clinch the diagnosis. A slaty or gray discoloration of macula associated with neovascularization and shallow detachment gives suspicion of a malignancy of choroid or retina. Exudates in the macular area may be found in diabetic retinopathy, hypertension, papilledema, neuroretinitis and age-related macular degeneration.

Retinal Vessels

The central retinal artery, a branch of ophthalmic artery, divides at the optic nerve head into a superior and an inferior papillary branch, from each of which come a nasal and a temporal branch either within or beyond the optic nerve head. All the four retinal branches continue to divide dichotomously into several branches spreading over the retina and reaching the ora serrata where they loop to form capillaries. The distribution and division of retinal branches show great variations, the nasal branches run more radially while the temporal sweep to avoid the macula. The retinal veins follow the retinal arteries lying mostly on the temporal side. The arteries can be distinguished from the veins by their color and caliber. The arteries are bright red in color, while the veins are purplish red. The arteries are narrower than the veins, the ratio of the caliber of an artery to a vein being 2:3.

In healthy young individuals, the walls of retinal vessels are transparent; during ophthalmoscopic examination a light reflex is obtained due to reflection from the arteries. When the wall of an artery is thickened due to arteriosclerosis or hypertension, the light reflex increases in brightness and width and the underlying blood column gives a burnished copper appearance (*copper-wire artery*). The excessive thickness of the wall causes total reflection of the light giving a silvery reflex (*silver-wire artery*). The vessels, particularly veins, may show striking tortuosity in diabetes mellitus, occlusion of the central retinal vein and blood dyscrasias. Sheathing of the vessels appears as white lines along the sides of the vessels. Generally, an artery crosses the vein lying anteriorly and does not obscure it, the two vessels share a common adventitia. In vascular sclerosis, the wall of the artery is thickened so that the vein is obscured. Compression, distal dilatation and displacement of the vein are some of the signs of vascular sclerosis found at the arteriovenous crossing.

Retinal venous pulsation may be found on or near the disk in about 80 percent of normal individuals due to transmission of the intraocular pressure. It can also be elicited by a slight pressure on the eyeball. The arterial pulsation is almost always pathological and seen in increased intraocular pressure with normal or lowered blood pressure.

General Fundus

The appearance of fundus varies considerably from race to race depending on the degree of pigment in the retinal pigment epithelium. Normally, the fundus has a uniform red color. In black people the fundus is dark red, but in albinos the fundus presents a characteristic appearance due to lack of pigments. The choroidal vessels are clearly seen and the white sclera shines through the space between them. Occasionally, the pigment in the retinal pigment epithelium is deficient but the pigment in the choroid is marked; pigmented, polygonal areas are seen separating the choroidal vessels, such a fundus is known as *tigroid* or *tessellated fundus* (Fig. 9.44).

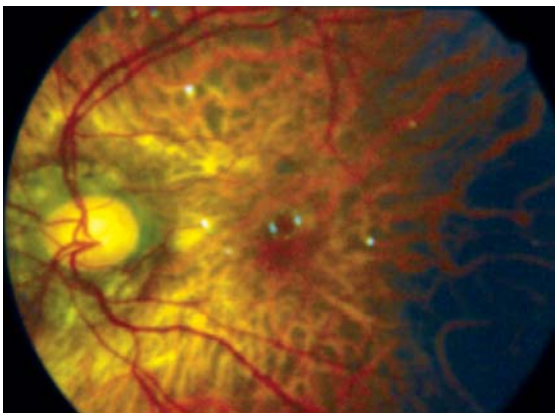


Fig. 9.44: Tessellated fundus

In high refractive errors, the appearance of the fundus is characteristic—shimmering reflex or watered-silk appearance seen in high hypermetropia, and myopic crescent and chorioretinal degeneration in high myopia.

In pathological states, the fundus may present hemorrhages, exudates, drusen, opaque nerve fibers, microaneurysms, neovascularization, tumors and retinal detachment.

Hemorrhages

Hemorrhages (Fig.9.45) may be preretinal or intraretinal. The *preretinal* (*subhyaloid*) *hemorrhage* is a large, round or hemispherical hemorrhage near the macula, while *intraretinal hemorrhages* may be found in the nerve fiber layer where they assume flame-shaped appearance or in the deeper layers appearing as round or irregular.

Exudates

The retinal exudates may be soft or hard (Fig. 9.45). Soft exudates or *cotton-wool spots* have fluffy, indistinct margins, but the *hard exudates* are small, discrete, waxy looking with crenated margins.

Drusen

Drusen (Fig. 9.46) or colloid bodies are numerous, minute, yellow lesions at the level of retinal pigment epithelium often found at the posterior pole due to deposition of abnormal basement-like material on Bruch's membrane.

Myelinated Nerve Fibers

Opaque or myelinated nerve fibers are due to extension of myelination beyond the lamina cribrosa in postnatal period. They are characterized by white, feathery patches usually continuous with the disk (Fig. 9.47) and, occasionally, isolated in the retina.

Abnormal Pigmentation

Pathological pigmentation in the fundus may appear either in a pepper and salt form, or in a bone corpuscular form as seen in retinitis pigmentosa.

Chorioretinal Lesions

Active or healed patches of chorioretinitis (Fig. 9.48) may be found near the disk (juxtapapillary), at the macula (central) or towards the periphery (anterior).

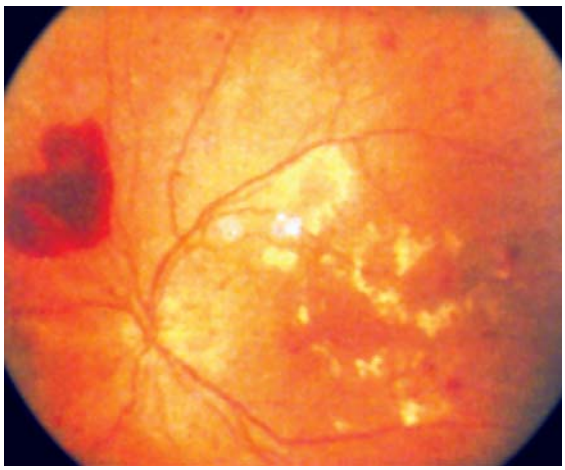


Fig. 9.45: Hemorrhages and exudates

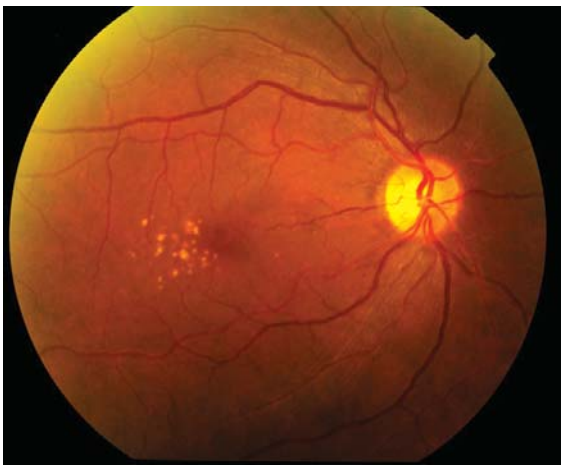


Fig. 9.46: Drusen

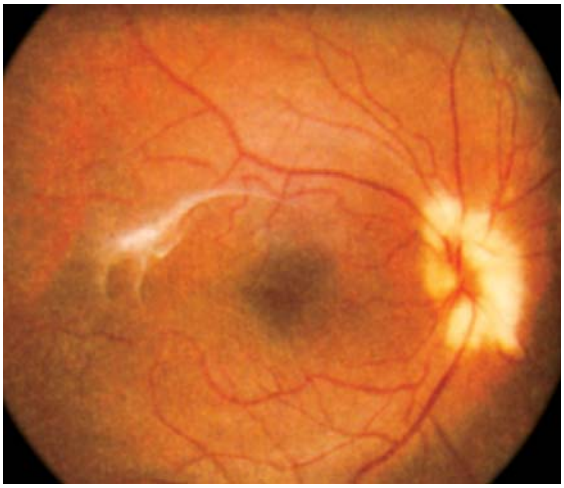


Fig. 9.47: Myelinated nerve fibers

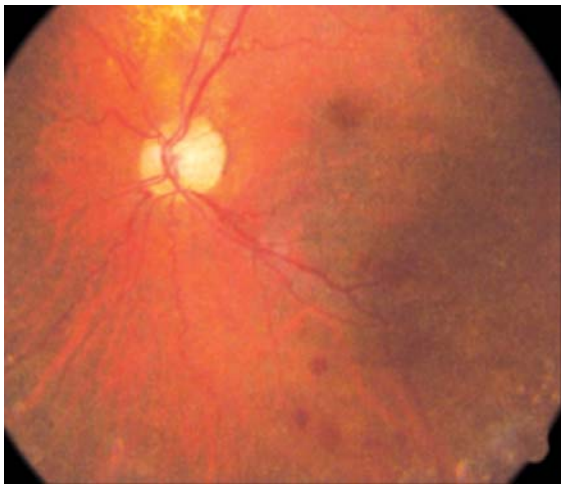


Fig. 9.49: Microaneurysms

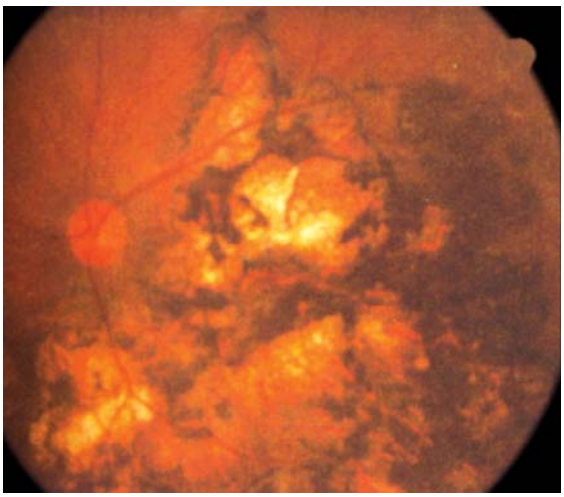


Fig. 9.48: Healed patches of chorioretinitis

Microaneurysms

Microaneurysms (Fig. 9.49) are multiple, pin-head dilatations of venules at the posterior pole and are found in diabetes mellitus and retinal vein occlusion.

Neovascularization

Fresh vascular channels may be formed in the retina to compensate for hypoxia. They often get ruptured and lead to hemorrhages.

Tumors

Both benign and malignant, pigmented or nonpigmented, tumors may arise from the retina. Large tumors frequently change the fundus reflex either due to pigmentary changes or associated retinal detachment.

Retinal Detachment

Retinal detachment obscures the view of the fundus partially or completely depending upon its extent. Breaks (atrophic or horse-shoe) or dialysis (Fig. 9.50) are frequently associated with primary type of detachment. A secondary detachment of retina is often seen in intraocular tumor (malignant melanoma) or Vogt-Koyanagi-Harada syndrome. Tractional detachment of retina results from fibrovascular proliferation associated with diabetic retinopathy or Eales' disease.

Fundus lesions, especially retinal breaks and neoplasms, require accurate measurements and localization. The measurement is expressed either in terms of disk-diameters or by actual measurement with the help of a scale or graticule attached to an ophthalmoscope. Papilledema and cupping of the disk can be appreciated by visualizing the blood vessels emerging from the disk. Then the

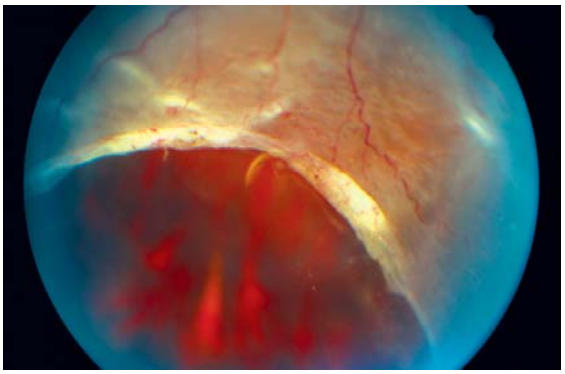


Fig. 9.50: Retinal detachment with retinal dialysis

extent of elevation or depression is directly measured by keeping the ophthalmoscope at 15.7 mm in front of the cornea and focusing the surface of the optic disk. A difference of focusing of 3 D between the blood vessels and the surface of the disk indicates a swelling or cupping of approximately 1 mm.

For the purpose of management, the distance of the lesion is measured either from the limbus or from the ora serrata after assessing the meridian in which it lies (o' clock position).

Examination of Retinal Functions

Each eye must be examined separately for its retinal functions. The retinal functions consist of the form sense or visual acuity, the color sense and the field of vision.

Visual Acuity

Visual acuity applies to central vision only and is tested both for distance and near. The visual acuity for distance (distant vision) is usually tested by means of Snellen's test-types.

The Snellen's test-types are constructed on the principle that two distant points can be visible as separate only when the minimum angle subtended by them at the nodal point of the eye is 1 minute. This forms the standard of normal visual acuity. The visual acuity depends upon the resolving power of the eye and varies with the wavelength of the light and size of the pupillary aperture.

The Snellen's test-types consist of a series of letters arranged in lines (see Fig. 7.9). The size of the letters gradually diminishes from above downwards and a numerical number is written underneath each line. Each letter is so designed that it fits in a square the sides of which are five times the breadth of the constituent lines. Therefore, at a given distance, the letter subtends an angle of 5 minutes at the nodal point of the eye (Fig. 9.51). The top letter of the chart subtends a 5 minute angle at the nodal point of the eye from a distance of 60 meters. The letters in the subsequent lines subtend same angle if they are 36, 24, 18, 12, 9 and 6 meters away from the eye. The chart should be well-illuminated and illumination should not fall below 20 foot candles. Some increase in visual acuity is noted with increase in illumination up to a certain point of brightness.

For recording the visual acuity the patient should be seated at a distance of 6 meters from the chart as the rays of light are practically parallel from this distance and accommodation is negligible. When the space in the room is limited, the test-types may be seen after being reflected from a plane mirror kept at a distance of 3 meters from the patient. The patient is asked to read the test-types after covering one eye either by a cardboard or by palm of the hand. The visual acuity is expressed as a fraction, the numerator of which is the distance of the chart from the patient (6 meters)

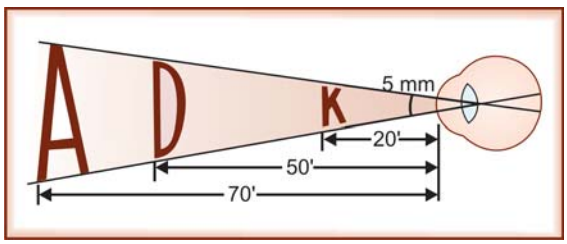


Fig. 9.51: The letters of the distant vision test types subtend a visual angle of 5 minutes at nodal point

and the denominator is the numerical number written underneath the line up to which the patient can read. For example if a patient can only read the top letter, his visual acuity is recorded as 6/60. In fact, a normal person ought to have read the letter from a distance of 60 meters. When patient reads the second, third, fourth, fifth, sixth and seventh lines the visual acuity of the patient is recorded as 6/36, 6/24, 6/18, 6/12, 6/9 and 6/6, respectively.

Normally, a person can read the line marked 6 and the visual acuity is expressed as 6/6. When the top letter cannot be read, the patient is asked to move towards the chart and if he reads the top letter from 3 meters distance, the visual acuity is recorded as 3/60. If the patient cannot appreciate the top letter even from a distance of 1/2 meter then the distance in feet or inches is recorded at which finger counting (FC) is possible. When he fails to count the fingers, see whether he appreciates the movements of the hand (HM). In absence of recognition of hand movements, perception of light (PL) should be tested.

The examination should be repeated for the fellow eye. In illiterate person 'E' test-types or Landolt's broken rings should be used. The testing of visual acuity in young children is a painstaking procedure requiring the use of pictures of different objects, circles and dots, and letters and numerals. For patients using glasses, the visual acuity should be recorded without glasses as well as with correction.

The Snellen's test-types measure vision at 100% contrast and are not sensitive to record subtle defects in visual function. Following sensitive tests may be employed to uncover these deficits.

Contrast Sensitivity Test

The contrast sensitivity test is used to record the visual acuity at various spatial frequencies and contrast levels. The patient is asked to identify letters from an array of equal-sized letters of diminishing contrast relative to white background (Fig. 9.52). The contrast sensitivity may be impaired in pseudophakic patients with 6/6 visual acuity recorded with Snellen's chart. Defective contrast sensitivity may be found in patients with glaucoma, lenticular opacities, amblyopia, optic nerve lesions, and refractive errors.

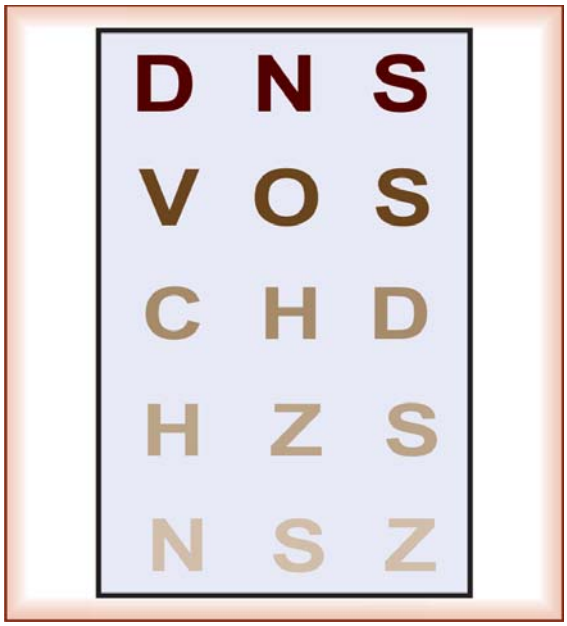


Fig. 9.52: Contrast sensitivity chart

Potential Acuity Tests

The pinhole test, the potential acuity meter test and the laser interferometer test are utilized to distinguish between visual dysfunction caused by aberrations of optical media (refractive errors, corneal surface defects and cataract) and organic lesions of optic nerve and retina.

Pinhole test: When viewing through a pinhole or multiple pinholes in a disk improves the subnormal vision, then either the refractive error or defects in the ocular media are responsible for the visual defect.

Potential acuity meter (PAM) test: The patient sees a miniaturized Snellen's chart projected onto the retina from a box mounted on the slit-lamp. The small image of the chart is often able to pass through the defects in media and in patients with refractive errors. The test provides accurate results except in cystoid macular edema where it over estimates the vision.

L*aser interferometer test*: It is based on the phenomenon of interference. Two pin-points of a laser light are focused on the anterior surface of lens. As the light enters into the eye, these points interfere with each other and form light and dark fringe patterns on the retina. A rough estimate of visual acuity can be made by changing the distance between two pin-points resulting in the alteration of fringe pattern.

Near Vision

The near vision is tested by using test-types in print. Jaeger's test-types or Roman test-types are in common use. In Jaeger's test types, a series of different sizes of print types are arranged in increasing order and arbitrarily marked 1, 2, 3, 4, 5, 6 and 7. The near vision is tested in good illumination preferably in day light. The patient is asked to read the chart kept at a distance of 25 cm. Generally, a person with normal vision and accommodation reads the smallest types easily. If unable to read the smallest types, the types which the patient can read should be noted. Patients with high hypermetropia, presbyopia or anomalies of accommodation have defective near vision.

Malingering

With the rapid industrialization and increasing stress and strain of life, many persons purposely pretend visual defect (even sudden loss of vision) without obvious organic lesions with the hope of gaining compensation or other advantages. The blindness may be feigned either in one eye or in both. A rough assessment of bilateral blindness can be made by watching the behavior and movement of the patient as well as pupillary reactions. However, if the defect is confined to one eye, it can be verified by the following tests.

1. Place a weak convex lens before the so-called blind eye and + 10 D spherical lens before the good eye and ask the patient to read the Snellen's chart. If he/she reads the letter the patient is malingering.
2. Place a 10 D base-out prism in front of the blind eye. A malingerer will move the eye inward to avoid diplopia.
3. The patient after wearing a pair of red-green glasses is asked to read the word 'FRIEND' written in alternate green and red letters on the vision drum (FRIEND test). The reading of the word exposes the malingerer.

Color Vision

A normal human being can perceive the primary colors—red, green and blue, and their shades. Certain occupations such as railways, navy, airforce require good color perception. Total color blindness is a rarity, however, defects in perception of colors are seen. The color vision is tested by various methods: Ishihara's pseudo-isochromatic chart, Edridge-Green lantern test, Holmgren's wool test, Nagel's anomaloscope and Farnsworth-Munsell-100-hue or D-15 test.

1. The *Ishihara's charts* (Fig. 9.53) are commonly used to determine the patient's ability to perceive colors. The charts are made up of dots

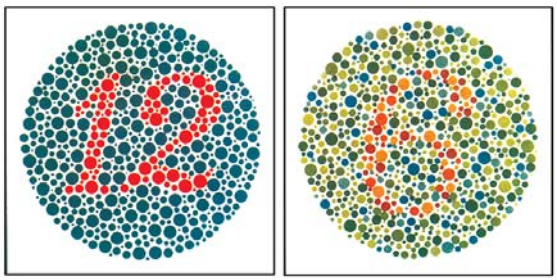


Fig. 9.53: Ishihara's chart plates

of primary colors printed on a background of similar dots of confusing colors. The dots are arranged to form numbers or certain patterns. Under 30-50 foot candles of illumination, the charts are presented to the patient at a convenient distance of 30 cm. He is asked to read the number or move a pointer over the pattern. A person having defective color vision is unable to read the number correctly or follow the contour of the pattern. The type of color deficiency can be diagnosed according to the chart used.

2. In *Edridge-Green lantern test* the subject is asked to name various colors shown from a lantern and a rough estimate is made depending upon the mistakes he makes.
3. In *Holmgren's wools test* different samples of colored wools are presented to the subject and he is asked to match color from a heap of colored wools. A color deficient subject will make wrong matches.
4. In *Nagel's anomaloscope* the subject looks through a telescope and is asked to match the color of the one-half of the illuminated disk with the other half by turning the knobs.
5. *Fransworth-Munsell-100-hue test* consists of 85 colored tablets and the subject is asked to arrange the tablets in a sequence. A normal individual will arrange them with minimum errors while the color deficient will commit mistakes in those parts of spectrum complementary to his color defect. The score is plotted on a chart. The test is very sensitive but time consuming. Hence *Fransworth D-15 test*, which is more rapid but may miss mild color deficiencies, is employed.

The color deficiency may be congenital or acquired. The red-green color deficiency is the commonest and is found in about 3-4 percent of male and 0.4 percent female population. The acquired color defects may occur in macular, retinal and optic nerve diseases. The retinal diseases show blue-green deficiency while, optic nerve diseases show a relative red-green deficiency.

Field of Vision

When the eye fixes its gaze on an object, the entire area which can be seen around the object is known as the *field of vision*. The field of vision can be tested either by confrontation test or by the use of perimeter.

Confrontation test is a rough method of assessment in which the patient's visual field is compared with that of the examiner. It is, therefore, essential that the latter should have a normal visual field. The patient sits at a distance of 2 feet from the examiner and is asked to close his left eye and fix his right eye on examiner's left eye. The examiner shuts his right eye and moves his finger from the periphery to the seeing area between him and the patient. The patient is instructed to tell as soon as he sees the finger. The finger is moved along the various meridians and thus a rough assessment is made about the visual field of the right eye. The test should be repeated for the left eye.

Perimetry is a technique for recording the visual fields with the help of an instrument called *perimeter*. Both central and peripheral fields of vision can be recorded with perimeters. The screen of perimeter may be either an arc or a bowl, the latter is preferred. Perimetry is essentially of two types—*kinetic* and *static*.

***Kinetic perimetry*:** A test object is moved from a non-seeing area, and the point at which it is first seen is recorded while patient fixates his eye. The test object should always be moved from the periphery towards fixation or from the center of the scotoma. Lister's perimeter and Goldmann's perimeter are used for this purpose.

Lister's perimeter (Fig. 9.54) is a simple instrument for recording the extent of peripheral visual field. It consists of a metallic semi-circular arc which can

Fig. 9.54: Lister's perimeter

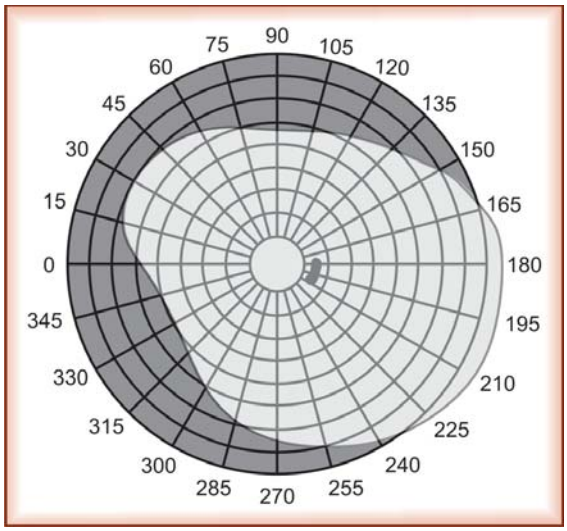


Fig. 9.55: Normal visual field of the right eye

be rotated in different meridians and has a self-registering recording device. Each eye should be separately tested. The patient keeps the chin on the chin-rest and fixes the eye on a white dot 330 mm away from the eye. A test object (3 mm) white or colored is gradually moved from the periphery of the arc towards the center. The patient taps the finger or a coin as soon as the object is sighted. From the recording device a perforation is made on the attached chart. Then the arc of the perimeter is moved by 30° and again the procedure is repeated. It is advisable to take 12 readings to complete the circle. The details of the size and color of the object, its distance, and date of recording should be noted on the chart.

The normal visual field for white object (5 mm) extends upwards 60°, downwards 70°, inwards 60° and outwards 90° (Fig. 9.55). For colored objects, the field is largest for blue and gradually decreases in descending order for yellow, red and green.

Goldmann's perimeter has a bowl of 300 mm radius which extends 95° to each side of fixation. It can be used to record both kinetic and static visual fields. The target is moved onto the bowl from black to white side and the points at which the patient fails to recognize it are noted. For static perimetry record, a self-illuminating target with an on-off switch is momentarily presented and the points at which the patient fails to perceive them are noted. The reliability and reproducibility of Goldmann's perimetry is greater than Lister's perimetry.

Campimetry (scotometry) is a type of kinetic perimetry which enables the examiner to explore the central and paracentral areas (30°) of the visual field. Bjerrum's screen (Fig. 9.56) (at 1 or 2 meters distance) is used to chart the field. The patient fixates on a spot in the center of the screen with one eye, the other being occluded, and a target (1 to 10 mm in diameter) is moved from the periphery towards the center in various meridians. Initially, the blind spot is charted. It is located about 12°-15° temporal and slightly lower to the fixation spot, and measures 7.5° × 5.5°. Central or centrocecal scotoma (defect in visual field) is found in optic neuritis. Open-angle glaucoma causes characteristic visual field defects (Seidel's sign or arcuate or Bjerrum's scotoma).

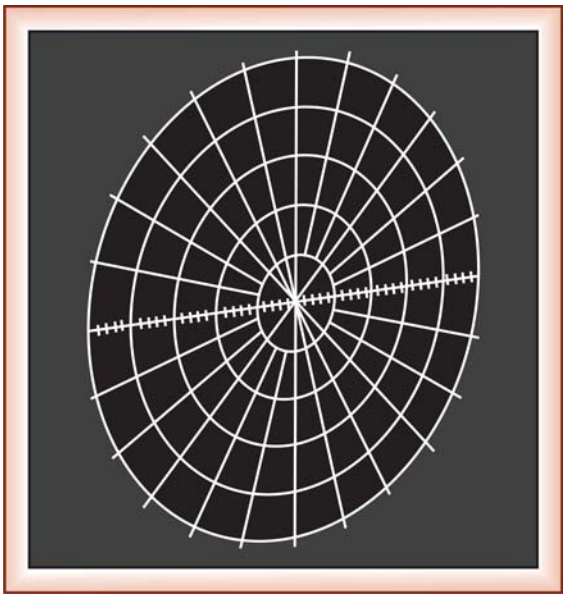


Fig. 9.56: Bjerrum's screen

***Static perimetry*:** It can be performed by Tubinger perimeter and automated perimeters besides Goldmann's perimeter.

Tubinger perimeter consists of a bowl-type screen and stationary test-targets with variable light intensity. The intensity of targets is increased from subthreshold level to a level at which the patient can recognize them (*threshold static perimetry*). A series of such points are plotted either along one meridian or in a circular manner. The technique is more sensitive than kinetic perimetry in detecting glaucomatous field defects.

Automated perimetry is widely used for the evaluation of visual field defects in ocular and neurological disorders. It is a computerized static visual field testing that estimates the retinal sensitivity at preselected locations (threshold perimetry). Retinal sensitivity is given by *threshold* which is defined as the stimulus sensitivity perceived 50 percent of time. The stimulus brighter than threshold intensity is called *suprathreshold* and dimmer one *infrathreshold*. The technique is quantifiable and repeatable. The results may be recorded and interpreted in gray-scale, and pattern-deviation plot (Figs 9.57 and 9.58) especially when one uses Humphrey field analyser (Fig. 9.59A) or Octopus (Fig. 9.59B).

Advantages of Automated Perimetry over Manual Perimetry

1. Automated perimetry is more accurate than manual perimetry.
2. It is quantifiable and repeatable.
3. Fixation in automated perimetry is constantly monitored.
4. The reliability of each test can be verified by false-positive and false-negative results immediately.
5. Automated perimetry provides statistical analysis and age-matched control to know how often such changes occur in normal population.

Limitations of Automated Perimetry

1. Automated perimetry is time consuming and some patients get tired and become uncooperative.
2. At least three consecutive examinations may be needed for establishing the diagnosis in the initial phase of the disease.
3. The perimeter is costly and a skilled technician may be needed to operate it.

Macular Function Tests

The visual acuity is maximum at the macula and, therefore, the integrity of the macula must be tested, especially when media are opaque. Following tests are employed.

1. *Cardboard test* is done in a dark room. One eye of the patient is covered and he/she is asked to look at a light source through a cardboard with two pinholes close together. If he/she can perceive two lights, the macula is normal.

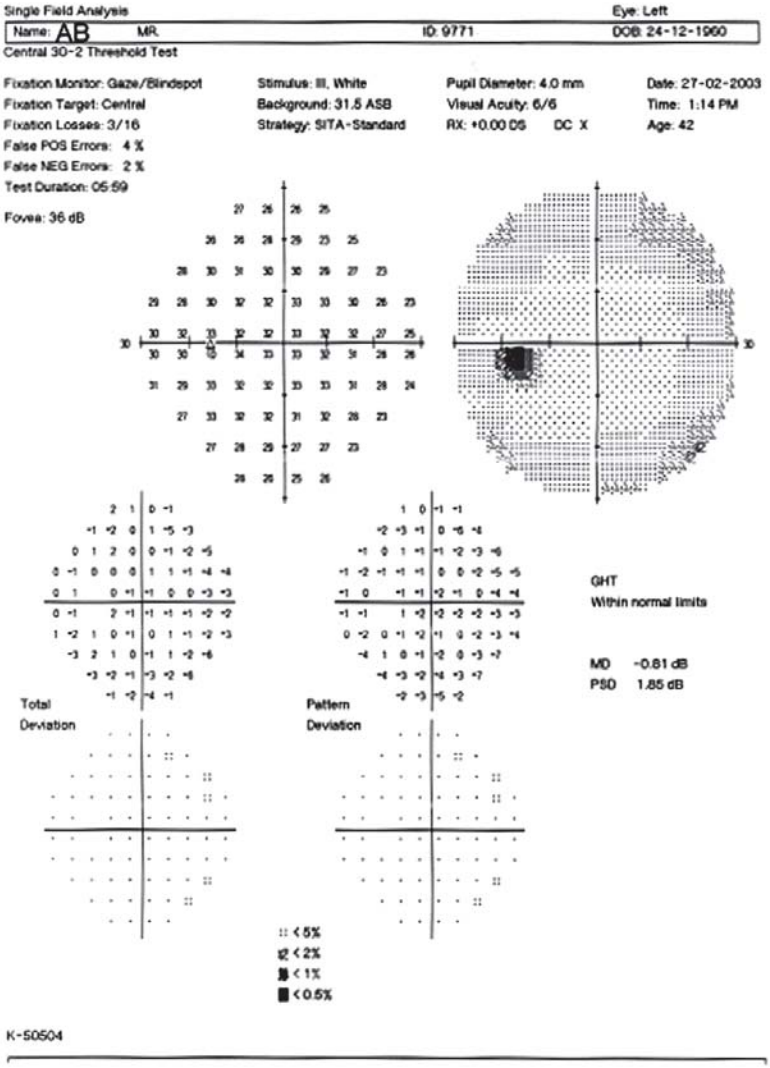


Fig. 9.57: Humphery visual field record (*Courtesy*: Dr D Sood, Glaucoma Imaging Centre, New Delhi)

2. *Maddox-rod test* is performed after covering one eye of the patient and he is asked to look at a light through a Maddox-rod. If a continuous red line is perceived, it indicates that the macula is normally functioning.
3. *Entoptic view test* is done by asking the patient to close his eyes, and the eyeball is massaged with the lighted bare bulb of a direct ophthalmoscope. He is then asked to describe what he sees. Generally, he sees clearly the entire vascular tree of the retina with a red central area. If macula is diseased, the central area will be dark rather than red and no blood vessels will be seen.
4. *Amsler grid* (Fig. 9.60) is used to detect macular disorders. The patient holds the chart at the

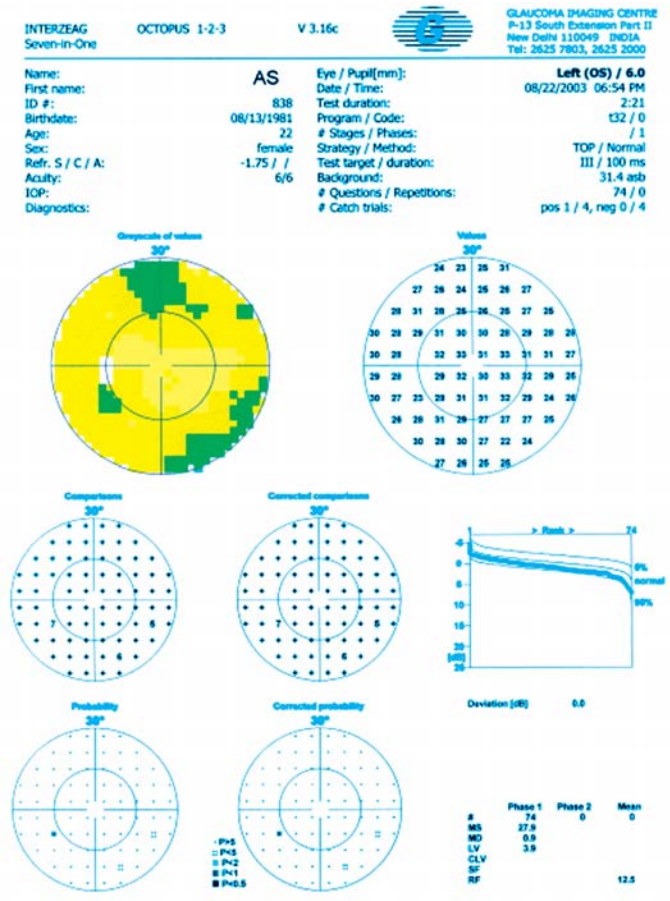
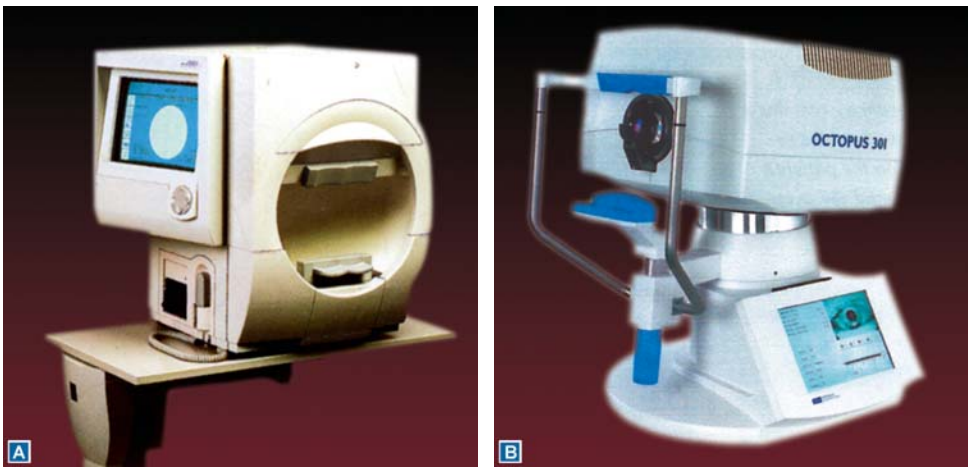


Fig. 9.58: Octopus visual field record (*Courtesy*: Dr D Sood, Glaucoma Imaging Centre, New Delhi)



Figs 9.59A and B: (A) Humphrey field analyser (Courtesy: Cart Zeiss, Bangalore), (B) Octopus automated perimeter (*Courtesy*: Biomedix Bangalore)

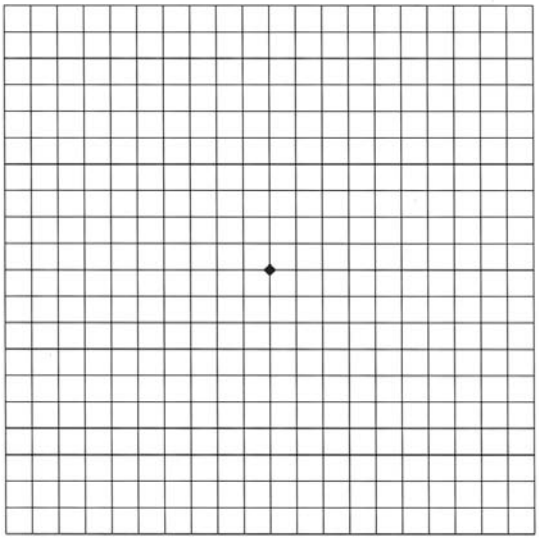


Fig. 9.60: Amsler grid chart

reading distance and keeps one eye closed. While fixing at the central dot, he is asked to outline any area of distortion or absence of grid.

Photostress Test

Photostress test is a subjective test that can be employed to diagnose the macular diseases. Cover one eye and record the baseline visual acuity. Then direct a beam of bright light of an indirect ophthalmoscope on the eye for 15 seconds and thereafter ask the patient to read the same line of the chart and note the recovery time. Repeat the test with the other eye. In normal individuals there is no difference in the recovery time. However, in patients with macular diseases, the photostress recovery time is prolonged significantly (normal is between 15 and 30 seconds).

Fluorescein Angiography

Fundus fluorescein angiography (FA) is of great diagnostic importance in vascular disorders of the retina and optic nerve such as diabetic retinopathy, retinal vein occlusion, Eales' disease, central serous retinopathy, age-related macular degeneration and optic neuropathy. Sterile sodium fluorescein (5 ml of a 10% or 3 ml of a 25% aqueous solution) is rapidly injected into the antecubital vein of the patient and serial photographs (Fig. 9.61) are taken with a fundus camera (Fig. 9.62) with paired filters. The excitation peak of fluorescein dye is about 490 nm and the emission peak is about 530 nm. Normally there are four overlapping phases in FA.

1. *Prearterial phase* is characterized by filling of the choroidal circulation.

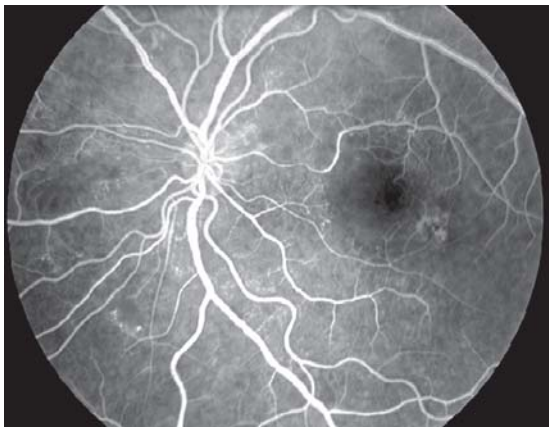


Fig. 9.61: Normal fluorescein angiogram

Fig. 9.62: Fundus camera
(*Courtesy*: Dr S Kanagami,Tokyo)

2. *Arterial phase* starts with the appearance of the dye in the retinal arteries.
3. *Arteriovenous phase* or *capillary phase* consists of filling of retinal arteries, capillary bed and early laminar flow in the veins.
4. *Venous phase* begins with venous filling and arterial emptying.

The fluorescein enters the choroidal circulation earlier (0.5 second) than the retinal. It appears first in the macular area and then spreads towards the periphery. The retinal pigment epithelium and the endothelium of the retinal vessels act as barriers to fluorescein and thus the dye remains confined to the intravascular space. However, in microaneurysms, neovascularization, arteriovenous shunts, defects in retinal pigment epithelium, prolonged venous stasis and inflammatory conditions of the retina and optic nerve, there occurs fluorescein leakage in the surrounding tissues. Retinal hemorrhages and pigmentation may obscure the underlying fluorescein.

Indocyanine Green Angiography

Indocyanine green angiography (ICGA) is an imaging technique used for studying the choroidal lesions. The indocyanine green dye has a longer excitation (805 nm) and emission (835 nm) wavelengths than the fluorescein. It can penetrate the retinal pigment epithelium (RPE) and the choroid without getting much absorbed by hemoglobin, melanin, exudates and lipid. Hence, the choroidal circulation and areas of neovascularization lying beneath the serous detachment of RPE (as occurs in age-related macular degeneration) show much better with ICGA than FA. The conjugation of ICG dye with proteins retains the dye within the choriocapillaris allowing distinct imaging of the choroid during angiography. The ICG angiograms may be recorded either using a modified fundus camera or scanning laser ophthalmoscope. The ICGA is quite useful in the diagnosis and management of age-related macular degeneration, central serous retinopathy, acute posterior multifocal placoid pigment epitheliopathy, Vogt-Koyanagi-Harada syndrome and serpiginous choroidopathy.

Electrophysiological Tests

There are two components of ocular potential, light-sensitive and light-insensitive. The light-sensitive component responds to changes in illumination and thus forms the basis of electrophysiological tests of retinal function.

Electroretinogram

Electroretinogram (ERG) measures the rapid changes in the resting ocular potential caused by exposure to a flash of light. The technique records the response between two electrodes, one being held in a corneal contact lens and the other placed on the forehead. The ERG consists of 4 waves: an initial negative a-wave, a large positive deflection, b-wave, slight positive deflection, c-wave, and an off response, d-wave. Both photopic (cone) and scotopic (rod) responses are obtained. Normally, b-wave response, arising from bipolar cell layer, is of 150 mv or over (Fig. 9.63). It is subnormal

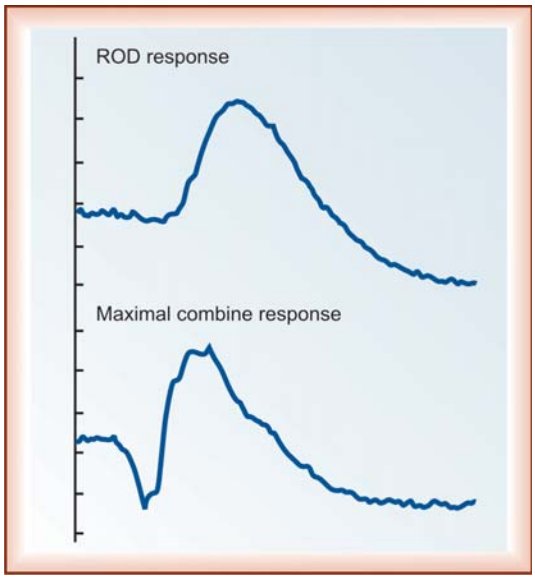


Fig. 9.63: Normal ERG
(Courtesy: Sankara Nethralaya, Chennai)

(less than 100 mv) in early primary retinal pigmentary degeneration even without ophthalmoscopic changes, and extinguishes in advanced pigmentary degeneration and cone dystrophy. Focal electroretinogram (FERG) can provide information about the macular function.

Electrooculogram

Electrooculogram (EOG) measures slow changes in the retinal potential during dark and light adaptation (Fig. 9.64). It is a whole retinal-response mediated through the rods. The electrodes are placed over the orbital margins near the medial and lateral canthi. The patient is asked to make side-to-side movements of the eye and recordings are made in dark and in light for 12 minutes each. The maximum height of light peak divided by minimum height of dark trough and multiplied by 100 gives the *Arden ratio*, which is normally 185 or above. The ratio is subnormal (less than 150) or flat (less than 125) in retinitis pigmentosa, degenerative myopia and macular dystrophies.

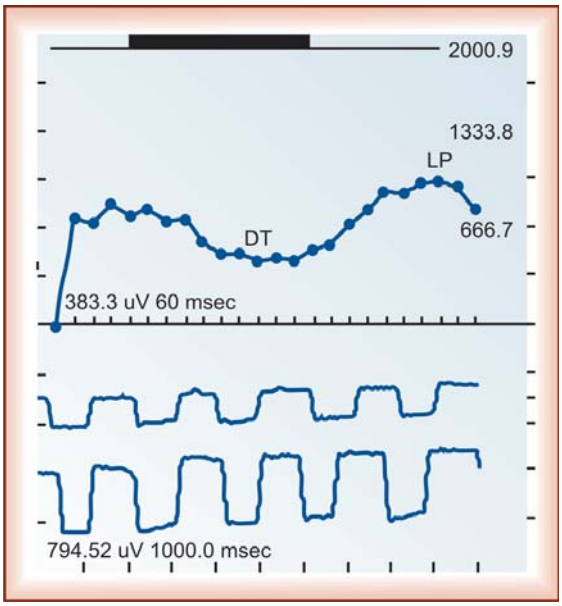


Fig. 9.64: Normal EOG (*Courtesy*: Sankara Nethralaya, Chennai)

Visual Evoked Potential

Visual evoked potential (VEP) is an important electrodiagnostic test to detect subclinical lesions of visual pathway. It is particularly useful in infants and uncooperative patients. When a flash of light strikes the retina, it evokes a volley of nerve impulses (Fig. 9.65). These impulses are transmitted along the visual pathway to the visual center. The velocity and quality of conduction are analyzed. Optic nerve lesions cause delayed conduction and reduced amplitude, while retrochiasmal lesions produce abnormal hemispheric responses.

Examination of the Orbit

Orbit is a rigid bony cage with only an anterior opening. Any increase in the orbital content is

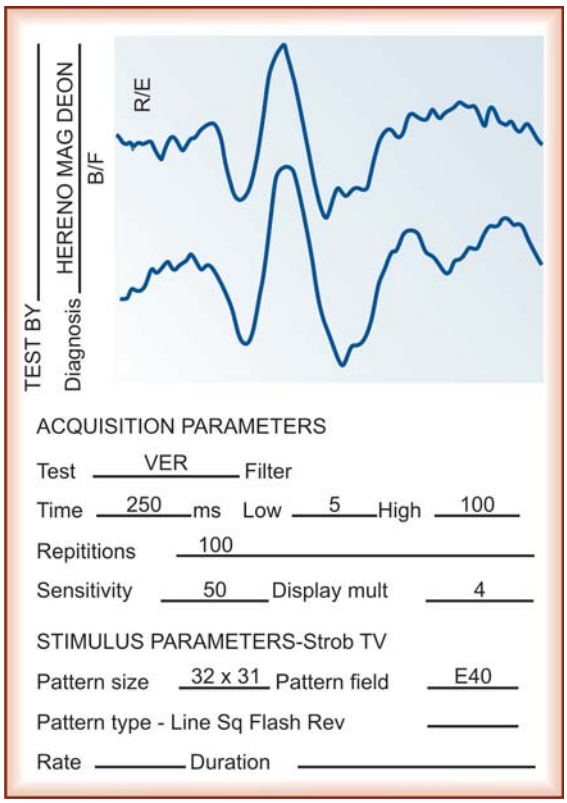


Fig. 9.65: Normal VEP

likely to displace or push the eyeball forwards—*proptosis* (exophthalmos). On the other hand, the senile atrophy of the orbital fat causes retraction of the eyeball in the orbit—*enophthalmos*. The proptosis may be due to retrobulbar tumor, orbital hemorrhage and orbital cellulitis. More often than not, displacement of the eyeball is accompanied with limitation of ocular movements and diplopia (double vision). Bilateral proptosis is caused by congenital shallowing of the orbital cavities and thyroid disorders. Pulsating proptosis may be found in carotid-cavernous fistula and vascular tumors.

The degree of proptosis can be accurately measured by exophthalmometry and the orbital lesions can be diagnosed by special techniques like ultrasonography, tomography and CT scan.

Exophthalmometry

Exophthalmometry is a method in which the degree of proptosis or the anterior protrusion of the eye is measured with the help of an instrument, the *exophthalmometer*. There are several types of exophthalmometer in use, but Hertel's exophthalmometer (Fig. 9.66) is often used in clinical practice. The patient stands facing the examiner. The concave parts of the instrument are placed on the lateral orbital margins. The distance between the lateral orbital walls is noted. For examining the right eye, the patient is asked to fix his right eye on the examiner's left eye. The image of the cornea coincides in the mirror with a scale (reading in mm). For the left eye, the patient is asked to fix his left eye on the examiner's right eye and the readings can be obtained in the same way. Normally, the readings of two eyes are more or less the same and range between 12 and 20 mm. A difference of more than 2 mm between the two eyes or any reading exceeding 20 mm should be considered abnormal.

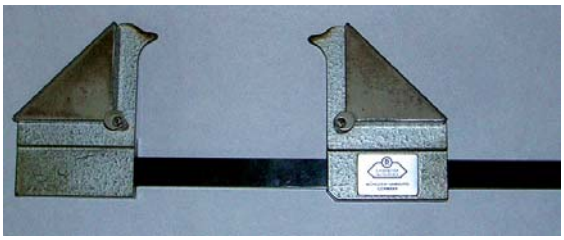


Fig. 9.66: Hertel's exophthalmometer

Ultrasonography (Echography)

A sound wave is referred as ultrasound if its frequency is over 20 kHz. It can penetrate ocular tissues regardless of the transparency. The ultrasound instrument consists of (i) a transmitter for electric energy, (ii) a transducer, and (iii) a cathode ray oscillograph. An ultrasound beam is directed into the eye under examination and a portion of the beam is reflected (where it encounters impedance) as echo. The echoes are converted to electrical impulses by transducer and displayed on a cathode ray tube. The echoes may be displayed as vertical deflection (*time amplitude* or *A-scan*) (Fig. 9.67) or bright spots and lines (*intensity modulated display* or *B-scan*) (Fig. 9.68) forming a picture of the eye and orbit like an X-ray photograph.

The Echorule 2 (Fig. 9.69), a portable A-scan biometer, measures the axial length of the eyeball

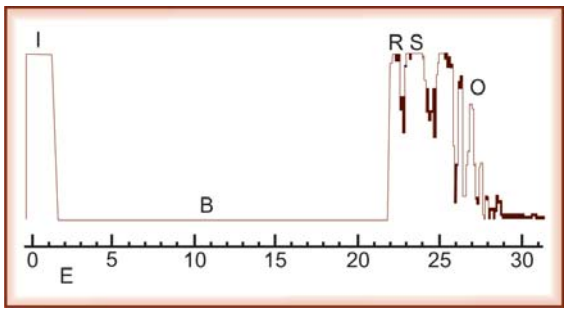


Fig. 9.67: Normal A-scan with sound beam bypassing lens; I—initial spike, B—baseline representing echo-free vitreous, R—retina, S—sclera, O—orbital soft tissues, E—electronic scale (*Courtesy*: Prof. Rajiv Nath, KGMC, Lucknow)

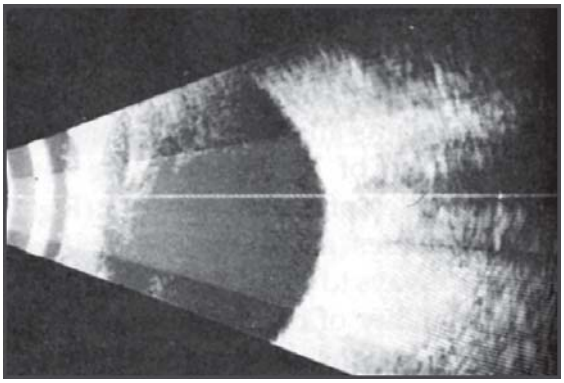


Fig. 9.68: Normal B-scan

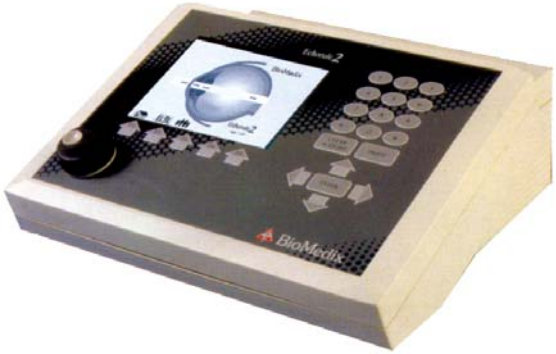


Fig. 9 69: Echorule 2 (Courtesy: Biomedix, Bangalore)

and facilitates calculation of the power of the intraocular lens (IOL) for implantation.

B-scan is of a greater value in the examination of orbit than the A-scan. It provides a very rapid and convenient examination of intraocular structures even in opaque media by using a transducer of about 10 MHz.

Ultrasound Biomicroscopy

Ultrasound biomicroscopy (UBM) is a method of procuring images of the anterior segment of the eye with high frequency ultrasound (40-100 MHz). The high frequency sound waves can image the anterior segment structures including the ciliary body with reasonably good resolution.

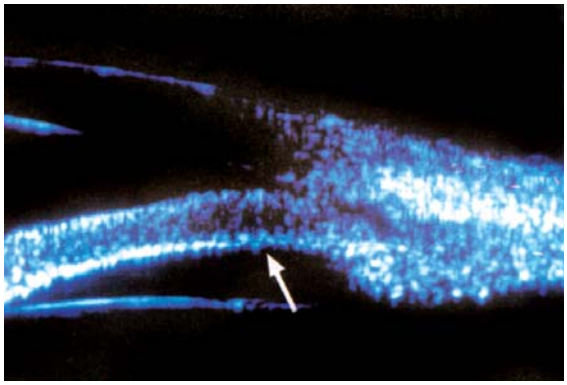


Fig. 9.70: UBM image showing the anterior convexity of the iris in primary angle-closure glaucoma (*Courtesy*: Drs K Prasad and SG Honavar, LVPEI, Hyderabad)

UBM is used for evaluating the anterior segment in eyes with corneal scars before penetrating keratoplasty, configuration of the angle of the anterior chamber, ciliary body tumors, localizing the anterior segment foreign body and in understanding the pathomechanisms of various types of glaucoma (Fig. 9.70).

Optical Coherence Tomography

Optical coherence tomography (OCT) is a technique which provides a cross-sectional image of the eye *in vivo* with a high resolution, similar to a histological section by light microscopy. OCT is very helpful in the diagnosis of macular hole (Fig. 9.71), macular edema, retinal detachment and nerve fiber layer defects in glaucoma. It is increasingly being used for establishing the diagnosis of macular hole and age-related macular degeneration.

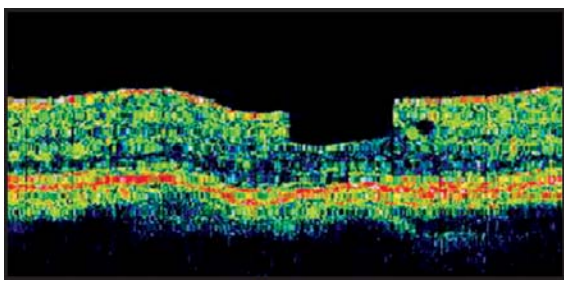


Fig. 9.71: OCT showing macular hole

Radiography

Radiographic techniques are commonly used either to diagnose the orbital lesions or to confirm the invasion of orbit by diseases of surrounding structures. The techniques may be *noninvasive* such as plain X-ray, tomography and computed tomography (CT) scan or *invasive*—arteriography, venography and pneumography, which are more or less replaced by color doppler imaging to determine the blood flow in carotid, ophthalmic or posterior ciliary artery.

Plain X-ray examination of the orbit is a frequently employed procedure. To obtain maximum information various views should be taken.

1. *Posteroanterior view* (Caldwell) demonstrates the superior orbital fissure, sphenoidal, ethmoidal and frontal sinuses and floor of the sella turcica.
2. *Water's view* provides details of orbital rim and floor of the orbit and maxillary sinus.
3. *Optic foramen view* (Fig. 9.72) should be obtained to compare the symmetry and size of the two foramina.

Tomography

The superimposition of paranasal sinuses, petrous part of the temporal bone and sphenoid bone upon the orbit does not allow proper evaluation of the orbit in plain X-ray. Tomographs of frontal, lateral and submentovertex projections are helpful in better evaluation of orbital lesions (particularly of the optic canal), as they provide two dimensional viewing.

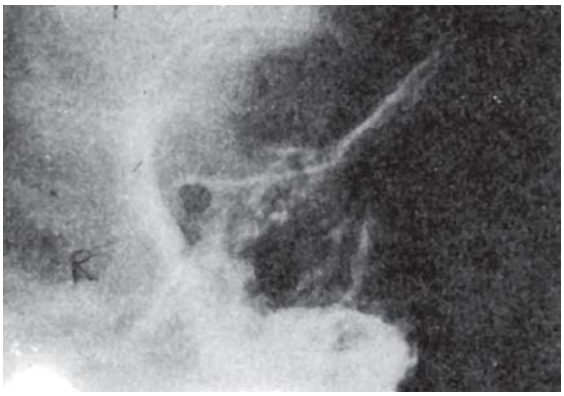


Fig. 9.72: Radiograph of optic foramen

Computed Tomography

The computerized tomography (CT), in axial or coronal plane uses thin X-ray beams to get the tissue density values, is a rapid and safe technique for examination of the orbit and brain (Fig. 9.73). CT scan allows accurate assessment of bony lesions, thickening of the optic nerve and extraocular muscles, orbital tumors or orbital invasion from intracranial or sinus malignancies.

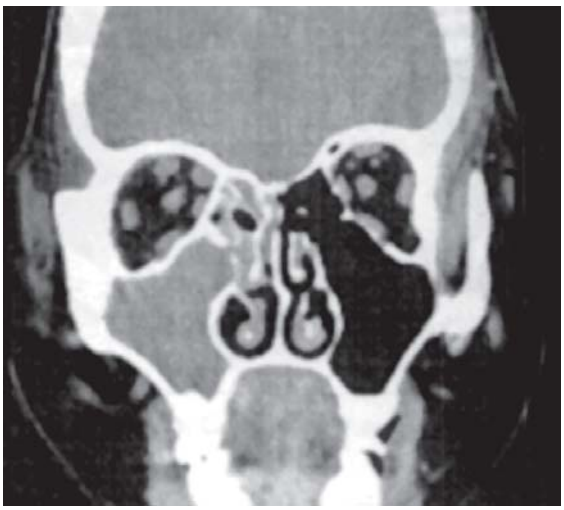


Fig. 9.73: CT scan cut at the level of orbit

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is a better imaging modality in the diagnosis of ocular and adnexal diseases as well as lesions of visual pathway. It is based on the electromagnetic properties of hydrogen nuclei which can be energized by pulses of radio frequency when placed in a strong magnetic field. The energy

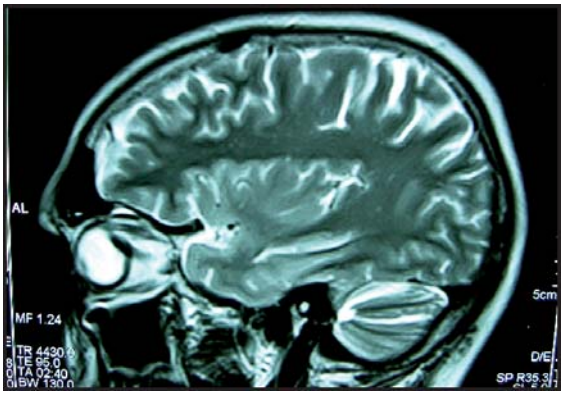


Fig. 9.74: Normal MRI

emitted during relaxation generates magnetic resonance images which are picked and analyzed by computers. MRI is, hence, free from the hazards of irradiation. It is preferred over CT scan for localization of a soft tissue lesion (Fig. 9.74).

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CHAPTER

10 Ocular Therapeutics

Various chemotherapeutic agents and antibiotics are widely used in prophylaxis and treatment of ocular infections. Indeed, they have amazingly changed the pattern of ocular diseases. Ophthalmia neonatorum, a blinding infection of the newborn, is virtually tamed and controlled with antibiotics. Trachoma, keratitis, corneal ulcer and ocular infections following intraocular surgery are being efficiently managed with the help of therapeutic agents. However, with indiscriminate use and abuse of these drugs, ocular toxic-allergic reactions and superinfections have cropped up. As each of the available drugs has one or more limitations in terms of efficacy and toxicity, research and clinical analysis of newer drugs or their synthetic derivatives are constantly being carried out in order to obtain maximal therapeutic benefit with minimal side effects.

CHOICE OF ANTIMICROBIAL AGENT

Optimal and judicious selection of antimicrobial agents for treating infectious conditions is a complex procedure that requires clinical judgement and a detailed knowledge of pharmacological and microbiological factors. When antimicrobial therapy is indicated, such a drug should be chosen that is selective for the infecting microorganism and which has the least potential to cause adverse reactions to the host. Initiation of rational antimicrobial therapy requires identification of the infecting organism by culture technique and testing its sensitivity to the available drugs. Since therapy may be required before the proper bacteriological identification, a simple gram's staining or KOH smear of the infected fluid will help to narrow the list of potential pathogens and permit a rational selection of the initial antimicrobial agent. The subsequent therapy will depend on the culture-sensitivity report because individual strains of microorganisms may vary widely in their sensitivity. If laboratory facilities are not available at all, therapy may be started on the basis of clinical diagnosis. This may only be provisional and it may later prove wrong, but the treatment chosen should be based on some explicit assumption as to the nature of the disease process. The next important factor to be considered is whether the drug is achieving inhibitory or bactericidal concentration at the site of infection. The location of an infection, to a large extent, dictates the choice of drug and the route of its administration. The minimum drug concentration achieved at the site should be at least equal to the minimal inhibitory concentration (MIC) for the infecting organism, although in most instances it should be about four to eight times of MIC.

MODES OF ADMINISTRATION

Most of the superficial ocular infections respond to topical therapy, while in severe intraocular infections it is essential to obtain quickly the adequate therapeutic concentrations of the antimicrobial agent in intraocular fluids and tissues. Under such circumstances, the agents are administered by subconjunctival, retrobulbar, peribulbar, intracameral or intravitreal route. More often than not, the drug is administered systemically by oral or parenteral route. Whenever a drug has to be given intravenously, the ophthalmologist has to choose either a slow continuous drip to obtain sustained concentration or periodic administration to produce intermittent peaks of high concentration.

Topical Therapy

Topically employed medicaments are used in the form of solution, suspension, gel and ointment. The latter obscures the vision, hence, convenient only for application at night. To avoid irritation to the eye, the solution should be isotonic and have a pH between 3.5 and 10.5. Generally, those antibiotics which are frequently used for the control of systemic infection should be avoided for topical use because of the danger of development of drug resistance.

The drug instilled in the conjunctival *cul-de-sac* is absorbed through the cornea. The concentration of a drug in the conjunctiva can be maintained for a longer period of time if the lower punctum is pressed by thumb to delay its nasal absorption.

The corneal epithelium forms the main barrier for intraocular drug penetration. The corneal stroma is permeable to all water soluble substances. The permeability of the corneal epithelium is increased if the solution has lower surface tension or is lipophilic or a wetting agent. The epithelial barrier is disrupted by the use of local anesthetic drop or by trauma.

A slow sustained release of the drug may be obtained by ocuserts placed in the upper or the lower fornix or by *drug impregnated contact lenses*. A collagen shield (contact lens) is an interesting device for antibiotic delivery. The shields are rehydrated in antibiotic solution prior to their placement on the eye. Over a period of time these collagen shields become gel-like and gel dissolved. The device acts as a drug reservoir and provides high level of drug in the cornea.

Subconjunctival Therapy

The superficial infection of the eye responds to topical therapy, but intraocular infections present a special problem because of differential ocular penetration of the drug. Since many drugs have poor intraocular penetration owing to their large molecular size, their adequate concentration can be achieved by means of subconjunctival injections, the choice being limited by their solubility and local tolerance. The subconjunctival route provides a speedy high concentration of the drug which may last for 2 to 3 days. Both antibiotics and steroids can be administered into the eye by subconjunctival route that by-passes the corneal epithelial barrier. Commonly used subconjunctival antibiotics for the treatment of corneal ulcer are listed in chapter on *Diseases of the Cornea* (Table 12.2).

Sub-Tenon Therapy

The sub-Tenon route is employed for a slow but sustained release of corticosteroids, especially in the management of pars planitis or recurring uveitis. Posterior sub-Tenon administration of methylprednisolone acetate or triamcinolone acetonide is useful for the management of chronic intraocular inflammations.

Retrobulbar Therapy

The retrobulbar route is used to anesthetize the eye for ocular surgery. Some ophthalmologists

choose this route for injecting steroids in the muscle-cone space for the management of posterior uveitis. The procedure is not without risk as it may cause perforation of the globe or damage to the optic nerve. Hence, peribulbar route is preferred.

Peribulbar Therapy

The peribulbar route is a safe route for administration of anesthetics, steroids or antibiotics. Peribulbar local anesthesia is recommended for almost all, except children and mentally disturbed uncooperative patients, posted for intraocular surgery. This route is also utilized for the management of thyroid ophthalmopathy and posterior uveitis.

Intracameral and Intravitreal Therapy

Injection into the eye either into the anterior chamber (intracameral) or in the vitreous (intravitreal) is employed in desperate cases. Intracameral injections of antibiotic are preferred when cultures are positive from the anterior chamber. Perhaps this route of administration has no real advantage over the subconjunctival injection of antibiotic. However, peroperatively intracameral pilocarpine or carbachol is used to achieve miosis, preservative-free lidocaine for relieving pain and 0.1% trypan blue or 0.5% indocyanine green for staining the lens capsule.

Intravitreal injections of antibiotics and antifungals are indicated in bacterial or fungal endophthalmitis, respectively. Intraocular injections are particularly suitable for the treatment of infected conditions which require surgery, for example, closure of a ruptured operative wound, repair of a lacerated wound and postoperative intraocular infection. The recommended intravitreal antibiotics and steroid injections with their doses are given in the chapter on *Diseases of the Uveal Tract* (Table 14.5).

Iontophoresis

Iontophoresis is a technique by which an electrolyte is given into the eye with passage of a galvanic current. The procedure is seldom used as it damages the corneal epithelium.

Systemic Route

Systemic administration of a drug by conventional route, oral or parenteral, has certain limitations because of impermeability of blood-aqueous barrier. The blood-aqueous barrier prevents the passage of large-sized molecules or water soluble compounds. Lipid soluble drugs (chloramphenicol, sulphonamides) can penetrate the barrier and diffuse into the aqueous humor in therapeutic concentration. Apart from lipid solubility, molecular weight, degree of protein binding, concentration of drug in blood and condition of blood-ocular barrier determine the intraocular penetration of a systemically administered drug.

CHEMOTHERAPY

Chemotherapy can be defined as the use of chemical compounds to destroy infective organisms without the destruction of their host. Dyes and heavy metal compounds like silver, arsenic, bismuth, mercury and antimony are active chemotherapeutic agents. But they should be used with caution and discretion because of their high toxicity.

Sulfonamides

A new era of chemotherapy was opened in 1935 with the introduction of prontocil, a sulfonamide. Chemically, the antimicrobial activity of sulfonamide depends on a free para-amino group and a direct link between the sulphur atom of the sulfonamide group with the benzene ring. Sulfonamides are usually administered orally, and depending upon their duration of action are classified as follows:

1. *Short-acting sulfonamides* (sulfanilamide, sulfamerazine, sulfacetamide, sulfixazole, sulfadimidine, sulfamethizole)
2. *Intermediate-acting sulfonamides* (sulfamethoxazole, sulfaphenazole)
3. *Long-acting sulfonamides* (sulfamethoxypyridazine, sulfadimethoxine, sulfamethoxine).

Sulfonamides are mainly bacteriostatic but in very high concentration they may act as bactericidal. They are effective against a number of gram-negative and gram-positive organisms and certain chlamydia, nocardia, actinomyces and toxoplasma infections. Sulfacetamide and sulfadiazine have good ocular penetration. A variety of microorganisms, *Pseudomonas pyocyanea, Corynebacterium diphtheriae, Salmonella* and *Mycobacterium tuberculosis* and anaerobic *Streptococci*, are not sensitive to sulfonamides. Adverse reactions to sulfonamides are not uncommon. Nausea, vomiting, fever and skin rash may develop. A severe exudative type of erythema associated with widespread lesions of skin and mucous membrane (Stevens-Johnson syndrome) has been noticed with long-acting sulfonamide therapy.

Sulfonamides may be administered either topically or systemically. Aqueous soluble sodium sulfacetamide (10%, 20%, or 30%) in drops or as ointment (6%) is used in conjunctival or corneal infections. The drug causes mild irritation. Systemic administration of sulfonamides gives high concentration in the aqueous as they are lipid soluble and pass the blood-aqueous barrier easily. Short-acting sulfonamides are administered in an average dose of 2 g initially followed by a six-hourly maintenance dose of 1 g. In children, the daily doses should be calculated on the basis of 150 mg/kg body weight, and be given in divided doses. In case of long-acting sulfonamides, an initial dose of 1 g is given and is followed by 0.5 g daily.

Trimethoprim-Sulfamethoxazole (Cotrimoxazole)

The introduction of trimethoprim with sulfamethoxazole is considered as an important advancement in chemotherapy. The antibacterial spectrum of trimethoprim is similar to that of sulfamethoxazole, but the former is usually 20 to 100 times more potent than the latter. All strains of *Streptococcus pneumoniae*, *C. diphtheriae* and *N. meningitidis* are sensitive to this combination. *Staphylococci*, *Streptococci*, *E. coli*, *Salmonella, Shigella* and *Pseudomonas pyocyanea* are also sensitive to cotrimoxazole. The combination acts on two steps of enzymatic pathway for the synthesis of tetrahydrofolic acid. The sulfonamide inhibits the incorporation of para-amino benzoic acid (PABA) into folic acid and trimethoprim inhibits the reduction of dihydrofolate to tetrahydrofolate. Further, trimethoprim is a highly selective inhibitor of dihydrofolate reductase of lower organisms. Cotrimoxazole is available in oral tablets containing 80 mg of trimethoprim and 400 mg of sulfamethoxazole. The usual adult dose is 2 tablets every twelve hours for 10 to 14 days for management of most of the ocular infections. The combination should be used with caution in children under twelve years of age and pregnant women.

Antibiotics

Antibiotics are substances obtained from microorganisms that in high dilution can inhibit the growth of other microorganisms. Majority of antibiotics are derived from fungi but some like bacitracin, polymyxin B and colistin are obtained from bacteria. Chloramphenicol is synthesized by chemical method. Antibiotics have a selective action on microorganisms, some affect primarily gram-positive bacteria, others inhibit gram-negative bacteria and still others inhibit only certain fungi, yeast or protozoa. Those inhibiting only one group of microorganisms are called *narrow-spectrum antibiotics*, while those inhibiting

both gram-positive and gram- negative bacteria, rickettsiae and chlamydia are termed *broad-spectrum antibiotics*.

Mechanisms of Action and Classification

There are several ways to classify antibiotic agents, however, the most common classification is based on their mode of action. An antibiotic may either be bactericidal or bacteriostatic. These agents can hit several targets in the bacteria namely, the cell wall, the cytoplasmic membrane, the ribosomes and the molecules involved in the transcription of genetic information (Table 10.1).

Penicillin

Penicillin, the most important of the antibiotics, was obtained from the mould *Penicillium notatum*. A variety of semisynthetic penicillins are now produced. Penicillin, a beta-lactam antibiotic, is widely used in the control of infection because of its wide range of bactericidal action. It is effective against cocci and gram-positive organisms, but gram-negative bacilli are relatively insensitive. Penicillin may be administered locally in the form of drops (5000 to 10000 units/ml), ointment (2000 units/g), powder or as subconjunctival injection. Drops should be instilled into the eye frequently (hourly or two hourly) to control acute conjunctivitis. Systemic administration of penicillin produces effective concentration in the tissues. Benzyl penicillin injection (500,000 units intramuscular eight hourly) or benzyl penicillin tablets (50000 to 500,000 units four hourly) produce therapeutic levels in the plasma. The drug is quite safe but some individuals are so sensitive to it

Table 10.1: Site of action of antibiotics

Site of action	*Antibiotics*	*Process interrupted*	*Type of activity*
1. Cell wall	Bacitracin, Cycloserine,	Mucopeptide synthesis of cell wall	Bactericidal
	Vancomycin, Cephalosporins, Penicillins, Methicillin, Cloxacillin, Nafcillin, Oxacillin, Ampicillin, Amoxycillin, Carbenicillin	Cell wall cross-linking	Bactericidal
2. Cell membrane	Amphotericin B	Membrane function and/ or integrity	Fungicidal
	Nystatin		Fungicidal
	Polymyxin B		Bactericidal
	Colistin A and B		Bactericidal
3. Ribosome 50-S	Chloramphenicol, Macrolides (Erythromycin, Oleandomycin, Spiramycin), Lincosamides (Lincomycin, Clindamycin)	Protein synthesis	Bacteriostatic
4. Ribosome 30-S	Aminoglycosides (Gentamicin, Kanamycin, Neomycin Streptomycin, Amikacin, Tobramycin, Spectinomycin) Tetracyclines	Wrong translation of genetic code, miscoding, inhibit initiation of protein synthesis by preventing the attachment of 30-S ribosome to m-RNA, protein synthesis	Bactericidal
5. Nucleic acid	Quinolone	Inhibits DNA gyrase	Pancidal
	Griseofulvin	DNA and RNA synthesis	Fungicidal
	Mitomycin C	DNA synthesis	Pancidal
	Rifamycin	RNA synthesis	Bactericidal

that just a skin test dose may cause a severe anaphylactic reaction. Furthermore, a number of organisms become resistant to it over a period of time. The semisynthetic penicillins may also be used.

Methicillin is effective against *Staphylococci* resistant to benzyl penicillin as it is not inactivated by penicillinase. It is acid labile and, hence, has to be administered by intramuscular (2 g 6 hourly) or intravenous (2 g dissolved in 50 ml of normal saline) route.

Cloxacillin has a weaker antimicrobial activity than benzyl penicillin but is 5 to 10 times more potent than methicillin. It is devoid of severe toxicity. Cloxacillin is administered orally in doses of 250 mg or 500 mg, six hourly depending on the severity of the infection. A derivative of cloxacillin, *dicloxacillin*, achieves blood levels twice that of cloxacillin on oral administration.

Ampicillin is found to be effective against gram-positive and gram-negative organisms. However, *Pseudomonas* and some strains of *Proteus* are resistant to the drug. It is ineffective against penicillin-resistant staphylococci. It can be administered by oral as well as intramuscular routes. The adult dose of ampicillin by both routes is 250 to 500 mg, six hourly. Rashes and diarrhoea may develop in some sensitive patients during therapy.

Amoxycillin is a broad-spectrum semisynthetic penicillin and administered orally in doses of 250 to 500 mg eight hourly. It has better absorption, lesser side-effects and longer half-life than ampicillin.

Cephalosporins

Cephalosporins, a class of β-lactam antibiotics, are derived from the mould *Cephalosporium acremonium*. They resemble penicillins in chemical structure and mechanism of action. They have high potency against gram-positive and gram-negative bacteria and penicillin-sensitive and penicillin-resistant *Staphylococci* and *Pneumococci*. Broad-spectrum activity, low incidence of resistance and fewer side effects are obvious advantages of cephalosporins over penicillins. Like penicillins, cephalosporins should also be given after a sensitivity test. Cephalosporins are classified into four generations depending on their activity.

First generation cephalosporins include cephazolin, cephalexin and cephalothin. Besides gram-positive cocci, they are also effective against *E. coli*, *Proteus* and *Klebsiella*. They can be given orally or by IM or IV route in the doses of 250 mg to 1 g, 8 hourly.

Second generation cephalosporins include cephamandole, cefaclor and cefoxitin. They show some additional activity against gram-negative and beta-lactamase-resistant organisms. Cefoxitin is effective against anaerobes. The second generation cephalosporins can be used in the doses of 0.75 g to 1.5 g, IM or IV, 8 hourly.

Third generation cephalosporins include cefotaxime, cefoperazone, ceftazidime, ceftriaxone and latamoxef. They are more active against gram-negative organisms including *Pseudomonas*. They are administered intravenously in the doses of 1-2 g per day.

Fourth generation cephalosporins include cefpirome and cefepime. The antibacterial activity of fourth generation cephalosporins resembles the third generation cephalosporins. Zwitterionic character of cefpirome permits better penetration through porin channels of gram-negative bacteria. The fourth generation cephalosporins are given in the doses of 1-2 g, IV, 12 hourly.

Macrolides

Macrolide antibiotics include erythromycin, azithromycin, roxithromycin and clarithromycin.

Erythromycin is a potent drug that exerts its antibacterial effect by inhibiting the bacterial

protein synthesis. It is effective against *Streptococci, Pneumococci, H. influenzae, N. gonorrhoeae, Treponema pallidum* and *Chlamydia trachomatis*. The drug may be administered orally (125-250 mg, 4 times a day) or intramuscularly (100 mg, twice a day).

Azithromycin is less potent than erythromycin against gram-positive bacteria but is more effective against gram-negative organisms. Azithromycin is quite useful against *Chlamydia trachomatis* and *Toxoplasma gondii*. It is administered as a single dose of 500-1500 mg which provides high tissue concentration.

Clarithromycin has greater antibacterial activity than erythromycin. It is administered twice daily. Both azithromycin and clarithromycin cause less gastrointestinal disturbances than erythromycin.

Lincosamide

Lincomycin is a bacteriostatic antibiotic having a spectrum of activity similar to that of erythromycin. It is administered orally in doses of 500 mg thrice daily. It can also be given intramuscularly or intravenously 600 mg, twelve hourly. The drug is employed in patients allergic to penicillin and erythromycin.

Clindamycin is a semisynthetic derivative of lincomycin which is bacteriostatic at low concentrations and bactericidal at higher ones. It is a superior drug to lincomycin, particularly in the treatment of infection due to *C. diphtheria, Nocardia, Actinomyces* and *Toxoplasma*. It is administered in doses of 150 to 450 mg, four times daily.

Vancomycin is usually given intravenously in the doses of 0.5 g, six hourly. The drug is used in penicillin and cephalosporin-resistant infections. It is bactericidal against gram-positive organisms and given intravitreally for microbial endophthalmitis.

Bacitracin is derived from *Bacillus subtilis* and it resembles penicillin in antimicrobial activity. It is not much absorbed orally and is very toxic if given parenterally. It is used topically in the control of superficial ocular infections as its intraocular penetration is poor. Generally, bacitracin drop or ointment (500-1000 units/ml) is used several times a day. Microorganisms seldom develop resistance to bacitracin.

Aminoglycoside Antibiotics

The aminoglycoside antibiotics are used to treat gram-negative infections. The common ones include streptomycin, kanamycin, gentamicin, tobramycin, amikacin, neomycin, and paramomycin. Framycetin, colistin and polymyxin B are the other antibiotics which are effective mainly against gram-negative organisms.

Streptomycin is obtained from *Streptomyces griseus*. It is water soluble and has a wide spectrum of antibacterial activity, especially against *M. tuberculosis, Shigella, E. coli, Proteus, Pseudomonas, H. influenzae, Brucella* and *Nocardia*. Topically, streptomycin 5000 units/ml may be used for the control of conjunctivitis, dacryocystitis or corneal ulcers. Streptomycin is administered intramuscularly in the dose of 1 g per day for the control of ocular tuberculosis supplemented with para-amino-salicylic acid (PAS) or isoniazid. It is a toxic drug and can damage the VIII cranial nerve.

Gentamicin is quite effective against *Pseudomonas, Proteus, E. coli* and *M. tuberculosis*. The minimum inhibitory concentration of gentamicin is lower than that of kanamycin. The drug can be administered topically (drops and ointment in the strength of 0.3%), subconjunctivally (10 mg), and systemically (80 mg, eight hourly) chiefly for the treatment of infection caused by *Pseudomonas* species. Parenteral drug therapy may produce vestibular damage and ototoxicity, particularly

in the presence of renal impairment. It should be avoided in pregnancy and in patients with compromized renal functions. It can be used intravitreally for the management of endophthalmitis. Gentamicin and carbenicillin act synergistically in the control of pseudomonas infection.

Tobramycin has antimicrobial spectrum and toxicity similar to that of gentamicin, but it is more effective than gentamicin against *Pseudomonas aeruginosa*. It is administered intramuscularly in the dose of 3.5 mg/kg/day, in three to four equally divided doses. The drug can be given topically as 0.3% eye drop or ointment.

Amikacin is a semisynthetic antibiotic having therapeutic indications and adverse reactions similar to those of gentamicin. However, it should be used in the control of gentamicin-resistant organisms. It has a synergistic action with vancomycin. The combination is used intravitreally for the treatment of endophthalmitis.

Neomycin, a polybasic water soluble antibiotic, is effective against a wide range of gram-positive and gram-negative organisms. It has a bactericidal action and is quite effective against *Acanthamoeba*. It is poorly absorbed on oral administration and its systemic use is not recommended because of high toxicity. Topically neomycin drops (0.5%) are administered to control superficial ocular infections. Frequently, neomycin is combined with other antibiotics to obtain broad-spectrum antimicrobial activity. A popular combination includes bacitracin, polymyxin B and neomycin. It is especially effective against *Proteus vulgaris*, which is resistant to most of the antimicrobial agents.

Framycetin is a water soluble antibiotic and has the antimicrobial spectrum and toxicity similar to those of neomycin. The use of the drug is restricted only to topical application. Framycetin sulfate (0.5%) drop or ointment is used several times a day to control superficial ocular infection, particularly caused by gram-positive and gram-negative bacilli including *Pseudomonas pyocyanea*.

Polypeptides

Colistin is a bactericidal antibiotic effective especially against many gram-negative organisms. It is used for the treatment of ocular infection caused by *Pseudomonas*.

Polymyxin B seems to be the least toxic antibiotic for topical use to control superficial ocular infections. The antimicrobial activity of polymyxin B is similar to that of colistin. For topical administration 0.1 to 0.25 percent sterile isotonic solution is employed several times a day. Occasionally, polymyxin B is injected subconjunctivally (5-10 mg) or intravitreally (0.1 mg) to control intractable ocular infections. To obtain broader antimicrobial activity, polymyxin B is combined with neomycin and bacitracin.

Rifamycin is obtained from *Streptomyces mediterranei*. It is effective against penicillin-resistant staphylococci and *M. tuberculosis*. Rifamycin is administered in the dose of 250 mg twice or thrice daily by intramuscular injection. A derivative of rifamycin is *rifampicin*.

Tetracyclines

Tetracyclines are a family of closely related antibiotics. The first, aureomycin (chlortetracycline), was discovered in 1947. This was followed by oxytetracycline (terramycin) and acromycin (tetracycline). Many semisynthetic tetracyclines, dimethyl-chlortetracycline (ledermycin), doxycycline, rolitetracycline (reverin) and minocycline, are used in clinical practice. The tetracyclines are bactericidal in high concentrations and bacteriostatic in clinically used concentrations. They act by interfering with protein synthesis. Resistance may develop due to decreased permeability of the antibiotics. Tetracyclines are broad-spectrum antibiotics, as in addition to their antimicrobial

activity against gram-positive and gram-negative organisms they inhibit the growth of certain *Actinomyces, Rickettsiae* and *Chlamydia trachomatis*. Tetracyclines are often used as ophthalmic drops or ointment in concentration of 0.5 to 1 percent to control superficial ocular infections. Oxytetracycline (250 mg capsule, 4 times a day) or doxycycline (200 mg daily for 2 days, then 100 mg daily) is given orally in the management of staphyloccocal lid infections, low grade anterior uveitis, corneal ulcer and florid trachoma, while reverin (500 mg twice daily IM or IV), oxytetracycline and tetracycline (100 mg IM at 6-8 hours intervals) are used to control severe ocular infections like orbital cellulitis, endophthalmitis or acute anterior uveitis. Tetracyclines are contraindicated in growing children and pregnant or nursing mothers.

Chloramphenicol

Chloramphenicol is a broad-spectrum antibiotic derived from *Streptomyces venezuelae*. The antibacterial spectrum of chloramphenicol is similar to that of tetracyclines. However, *S. typhi*, *H. influenzae* and *H. pertussis* are more susceptible and gram-positive cocci are less susceptible to chloramphenicol than to tetracycline. Chloramphenicol is lipid soluble, hence, the drug easily penetrates into the eye through the blood-aqueous barrier to provide therapeutic concentration. Chloramphenicol drop (0.5% in buffered solution) or ointment (1%) is used in the treatment of superficial ocular infections. It is administered orally in doses of 250 mg, 6 hourly. The drug should not be used indiscriminately as it may lead to agranulocytosis and gray-baby syndrome.

Fluoroquinolones

Fluoroquinolones are a family of antibacterial agents based on 4-quinolene nalidixic acid. They are bactericidal agents that selectively inhibit DNA gyrase (bacterial topoisomerase II). They are active against both gram-positive and gram-negative organisms and have variable activity against anaerobes. Fluoroquinolones are administered topically as well as systemically. Ciprofloxacin, norfloxacin, ofloxacin, lomefloxacin, pefloxacin gatifloxacin and moxifloxacin are commonly used. Systemic fluoroquinolones should be avoided in children owing to the risk of arthropathy.

Ciprofloxacin is a potent fluoroquinolone having broad-spectrum activity against *Staphylococcus*, *Streptococcus*, *Chlamydia trachomatis*, *Haemophilus influenzae* and *Neisseria gonorrhoeae*. The drug is quite effective against aminoglycoside-resistant strains of *P. aeruginosa*. Topical ciprofloxacin is used either in 0.3% solution or ointment form in the treatment of conjunctivitis and corneal ulcer. It is well-tolerated with practically no toxicity. Oral ciprofloxacin is administered in the doses of 250 mg, six hourly or 500 mg, twelve hourly and has good intraocular penetration (about 10% of serum concentration).

Norfloxacin has a more or less similar antimicrobial activity to ciprofloxacin, but is less effective against *P. aeruginosa*, *C. trachomatis* and *Streptococcus*. It is often used in the treatment of urinary infection. Both solution and ointment (0.3%) are available for topical ocular use. Topical norfloxacin has a greater corneal epithelial toxicity than topical ciprofloxacin. The recommended doses of oral norfloxacin for adults are 400 mg, twelve hourly.

Ofloxacin is intermediate between ciprofloxacin and norfloxacin in antimicrobial activity. The drug is available in 0.3 percent solution. The oral dose of ofloxacin is 200 to 400 mg, twelve hourly.

Pefloxacin is claimed to have a wider antibacterial spectrum, better ocular penetration and lesser chances of developing resistance as compared to other fluoroquinolones. Topical pefloxacin (0.3%) is effective against *P. aeruginosa*.

Gatifloxacin is a fourth-generation fluoroquinolone. It is effective against a wide range of gram-positive and gram-negative bacteria. It has a dual mode of action: inhibits both DNA gyrase and topoisomerase. It is found to be effective against some bacterial species resistant to ciprofloxacin and ofloxacin. Gatifloxacin penetrates well into the aqueous humor and the vitreous.

Levofloxacin is an advanced new generation fluoroquinolone. It is the pure S-enantiomer of ofloxacin. It has a higher binding affinity to DNA gyrase. Levofloxacin (0.5%) appears to have an expanded activity against gram-positive organisms as compared to the third-generation fluoroquinolone. It retains an excellent activity against gram-negative pathogens as well.

Moxifloxacin is a potent fourth generation fluoroquinolone which is quite effective in the management of ocular infections. Moxifloxacin ophthalmic solution (0.5%) is self-preserved at a near neutral pH of 6.8. It is more lipophilic and achieves better penetration into the cornea and ocular tissues than other fluoroquinolones. The drug has an enhanced activity against gram-positive organisms, atypical pathogens (Nocardia), and anaerobes while retaining a broad-spectrum coverage against gram-negative organisms. Moxifloxacin binds more effectively to topoisomerase II and IV resulting in a greater antimicrobial activity against gram-positive organisms. The drug shows a low risk of quinolone-related toxicity.

Fluoroquinolones when combined with cephazolin provide better protection against the organisms causing bacterial keratitis than either of the drugs used alone.

Adverse reactions of fluoroquinolones include gastrointestinal disturbances, headache, dizziness, insomnia, confusion, tremors, rashes and photosensitivity. Rarely tendonitis and tendon rupture may occur.

Antifungal Agents

A number of antifungal agents have been identified. Some of these drugs are effective topically as well as systemically. Broadly, these agents can be classified as sulfa drugs and silver preparations, polyene antibiotics, pyrimidine derivatives and azole derivatives.

Sulfa Drugs and Silver Preparations

Sulfa drugs both topically and orally were used for the control of ocular fungal infections in the past.

Silver sulfadiazine, an antimicrobial agent, derives synergistic effect from the combination of sulfadiazine and heavy metal silver. Silver atom binds to the DNA of the organism and prevents its replication. The drug is used topically as 1% cream 5 to 6 times in a day and is found to be effective in superficial fungal corneal ulcers. Silver sulfadiazine has been replaced with azole antifungal drugs for treating fungal ocular infections.

Polyene Antibiotics

Nystatin is effective against *Candida, Histoplasma, Trichophyton, Microsporum* and *Blastomyces*. The exact mode of action of this antibiotic is not known. The drug acts as a fungistatic in lower concentration. Nystatin ointment contains 100,000 units/gram and is employed locally in keratomycosis.

Natamycin (Pimaricin) is obtained from *Streptomyces natalensis* and shows activity against *Candida, Aspergillus, Trichophyton, Fusarium* and *Cephalosporium*. The antibiotic is mainly fungicidal and used as ophthalmic suspension (5%) to treat fungal corneal ulcer.

Amphotericin B is obtained from *Streptomyces nodosus*. It has a wide antifungal activity against both yeast and filamentous fungi. It is also effective against *Histoplasma, Cryptococcus, Sporotrichum* and

Blastomyces. As it is insoluble in water, amphotericin B is poorly absorbed from the gut. The drug is usually administered intravenously in 5 percent dextrose, initially in doses of 0.05 mg/kg and later increased to a total daily dose of 3-4 g. It is also used topically, in the form of drops and ointment (2.5%), as well as intravitreally (5-8 μg).

Azole Derivatives

Azole derivatives used as antifungal agents can be either imidazoles or triazoles. They interfere with the biosynthesis of ergosterol resulting in disruption of the fungal cell membrane.

Miconazole is an imidazole derivative used to treat ocular infections caused by yeast and filamentous fungi. It is effective against *Candida*, *Dermatophytes*, *Paracoccidioides* and some species of *Aspergillus*. Topically, it is used as 1 percent solution or 1 to 2 percent ointment several times in a day.

Ketoconazole has a wide spectrum of activity against *Candida*, *Dermatophytes*, *Cryptococcus*, *Histoplasma capsulatum* and *Blastomyces*. The drug can be used by both topical and oral routes. Topically, it is used as 1 percent solution and orally 200 mg, six to twelve hourly.

Fluconazole is a fluorinated triazole with a limited antifungal spectrum. It is effective against *Candida* and *Cryptococcus* infections. It can be administered orally, in doses of 400 to 800 mg/day for several weeks, topically as 0.2% eye drop or intravitreally (1 mg/0.05 ml).

Itraconazole is effective against *Aspergillus*. It is administered either orally, 100-400 mg/day, or topically, as 1% eye drop.

Antiviral Agents

Antiviral agents are selectively active against either RNA or DNA viruses.

Idoxuridine (5 iodo-2-deoxyuridine) is structurally related to thymidine and acts by competing with thymidine in the biosynthesis of DNA. The drug is topically used as 0.1 percent aqueous solution every hour during the day and 0.5 percent ointment at night in the treatment of herpes simplex keratitis and vaccinia keratitis. Epithelial lesions caused by herpes respond well to IDU therapy but the drug is ineffective in stromal herpes.

Vidarabine or *adenine arabinoside* (Vira-A or Ara-A) was first utilized in cancer chemotherapy but later found to be more active as an antiviral agent. It is active against a number of DNA viruses and indicated for the treatment of herpes keratitis and viral keratoconjunctivitis. It is effective against herpes simplex superficial keratitis but ineffective in stromal disease. The drug acts by interfering with early steps in the synthesis of DNA. It is used as a 3 percent ophthalmic ointment 5 times a day.

Trifluorothymidine (TFT or *viroptic)* is topically a more potent antiviral drug than IDU and Ara-A in the treatment of herpetic keratitis. The agent is water soluble and used as 1 percent drops 5 to 10 times in a day.

Acycloguanosine (Acyclovir or *Zovirax)* is a potent antiviral drug which is selectively active against herpes. The DNA of herpes simplex virus produces thymidine kinase enzyme. Acyclovir is phosphorylated first to its monophosphate form in the presence of this enzyme and finally converted into its active triphosphate form by host cell enzyme. The active form has an affinity for viral DNA polymerase and, thus, no new viral DNA is synthesized. Acyclovir is a drug of choice for the treatment of herpetic infection of eye, skin and genitals. It is more potent than IDU, TFT and Ara-A in herpetic infection of the cornea because it penetrates the intact corneal epithelium and stroma and produces therapeutic concentration in aqueous humor. The drug is used as 3 percent ointment 5 times a day. Acyclovir and Ara-A have synergistic effect. Acyclovir is also effective against cytomegalovirus which possesses a protein kinase that phosphorylates acyclovir. Oral acyclovir

800 mg 5 times a day for 10 days is beneficial in the treatment of acute keratouveitis due to herpes zoster ophthalmicus.

Penciclovir inhibits viral DNA polymerase and is available as 1% skin cream to be used 8 times a day for 4 days.

Famciclovir, a prodrug of penciclovir, is used orally 1 g 3 times a day, for 10 days in acute infections.

Valaciclovir, a l-valyl ester of acyclovir, is used orally 1 g twice a day for 10 days.

Interferon is a natural substance produced by the host cells in response to both DNA and RNA viral genome penetration. It protects the host from the virus and is active against both DNA and RNA viruses. Cellular DNA-dependent RNA synthesis is necessary for the antiviral activity of interferon. Interferon acts as a depressor for a cell specific protein that inhibits viral replication. Protein synthesis is, therefore, necessary for interferon action. Interferon is found to be effective in preventing the recurrence of herpetic infection.

Ganciclovir is a drug of choice for the treatment of cytomegalovirus (CMV) retinitis. The exact mode of action of the drug is not known, perhaps it does terminate new DNA synthesis. Ganciclovir is usually administered intravenously in the initial doses of 5 mg/kg twice daily for 2 weeks and is followed by long-term maintenance therapy (5 mg/kg once daily). Intravitreal implant of ganciclovir (4.5-6 mg) is available. Ganciclovir may cause bone marrow suppression.

Foscarnet, like ganciclovir, is used in the treatment of CMV retinitis in AIDS. It is also administered intravenously requiring an initial high dose induction therapy (20 mg/kg) followed by long-term maintenance therapy (0.16 mg/kg/min infusion). It is toxic to the kidney.

Zidovudine, a thymidine nucleoside analogue, has a selective action against human immunodeficiency virus (HIV). The drug is a nucleoside reverse transcriptase inhibitor that stops the viral replication. It can be administered orally in the doses of 1500 mg/kg/day but has a short half-life. It causes myelosuppression.

ANTI-INFLAMMATORY DRUGS

Corticosteroids

The corticosteroids are used effectively in several inflammatory and allergic ocular disorders. They have marked anti-inflammatory, antiallergic and immunosuppressive effects. The anti-inflammatory effects of steroids are nonspecific as they do not control the primary cause of inflammatory reaction. The effects are based on reducing the capillary permeability, maintenance of the integrity of cell membrane, stabilization of lysosome membrane and inhibiting lysozyme release from granulocytes. Antiallergic and anti-immunologic activities of steroids are due to suppression of cell-mediated hypersensitivity reaction and modification of immune responses. They, as such, do not cure the disease but temporarily block the exudative phase of inflammation. It is, therefore, on the cessation of steroid therapy that the disease may resume its natural course. A combination of chemotherapeutic agent with steroid, is therefore, recommended for the proper control of the disease.

Broadly, corticosteroids may be divided into three groups.

1. *Short-acting*—cortisone and hydrocortisone
2. *Intermediate-acting*—prednisolone, methyl prednisolone and triamcinolone, and
3. *Long-acting*—dexamethasone and betamethasone.

In ophthalmic practice, corticosteroids may be administered locally or systemically. For topical use betamethasone or dexamethasone phosphate drop (0.1%) or ointment (25 mg/g) is used. They

are indicated in the management of phlyctenular conjunctivitis, vernal keratoconjunctivitis, interstitial keratitis, rosacea keratitis, episcleritis and iritis. Subconjunctival injections of these corticosteroids in the doses of 1 mg/0.25 ml are administered biweekly in the treatment of acute/subacute anterior uveitis. Posterior uveitis is treated by peribulbar or posterior sub-Tenon injection of corticosteroids.

Prolonged local administration of corticosteroids may induce glaucoma in some patients. The use of medrysone ophthalmic suspension (1%) or fluorometholone 0.1 percent solution or loteprednol 0.5% 4 to 6 times daily is recommended in vernal keratoconjunctivitis, anterior uveitis and follow-up cases of ocular surgery because they have less pressure elevating effect. Higher dilutions (0.01-0.02%) of dexamethasone with acyclovir 3 percent topically can be instilled 3 to 4 times in viral stromal keratitis to clear the corneal haze.

In intraocular fulminating infections such as acute exudative iridocyclitis, choroiditis retinitis, and corticosteroids are also administered systemically either orally or parenterally. Synthetic glucocorticoids, prednisolone, dexamethasone, betamethasone and 6-methyl prednisolone, are rapidly absorbed when given by mouth. Prednisolone acetate is administered in doses of 1-2 mg/kg daily in divided doses. Prednisolone acetate suspension (25 mg/ml) is injected intramuscularly. Betamethasone or dexamethasone is given orally 5 mg daily (Table 10.2).

In acute infection, betamethasone or dexamethasone injection is administered intramuscularly or intravenously in doses of 4 to 10 mg daily and the therapy has to continue for a week or so, thereafter the patient is put on the maintenance dose. The drug must not be withdrawn abruptly as this may precipitate acute renal insufficiency. In practice the dose is gradually tapered off reducing it by 15 percent every fifth day.

Corticosteroids can cause toxic effects. The toxicity of steroids is related to dose and duration of therapy and individual susceptibility. The systemic administration of corticosteroids may aggravate diabetes, hypertension and tuberculosis, and produce bleeding from peptic ulcer. Myopathy, psychosis, osteoporosis, growth retardation and subcapsular cataract formation have been reported after prolonged systemic administration of steroids. Injudicious topical steroid therapy can lead to iatrogenic glaucoma, keratomycosis, herpetic keratitis and delayed wound healing.

Nonsteroidal Anti-inflammatory Drugs (NSAIDs)

Systemic NSAIDs

Nonsteroidal anti-inflammatory drugs are potent inhibitors of prostaglandin synthesis, mainly by blocking the enzyme cyclo-oxygenase. NSAIDs are indicated in the treatment of episcleritis, scleritis, anterior uveitis and cystoid macular edema (CME). The commonly used NSAIDs are aspirin (acetylsalicylic acid), ibuprofen, mefenemic acid, oxyphenbutazone, indomethacin and diclofenac sodium.

Aspirin is one of the most widely used drugs in the medical practice. It has antipyretic, analgesic and antirheumatic activities. It is administered orally 600 mg, 3 times a day.

Ibuprofen is orally administered in adult doses of 400 mg, 8 hourly to control ocular inflammation.

Mefenemic acid is a genuine antiphlogestic analgesic. It is administered in the doses of 250 mg, 4 to 6 hourly.

Oxyphenbutazone is a pyrazolone derivative and given orally 100 mg, 3 times a day.

Indomethacin is an indole derivative often used orally 25 mg, 3 times a day to prevent CME.

Table 10.2: Doses and administration routes of various steroid preparations

Steroids	Routes			
	Topical	*Subconjunctival*	*Oral*	*Parenteral*
1. Dexamethasone	0.1% soln. 0.1-0.5% oint.	1 mg	5 mg/day	5 mg/day
2. Betamethasone	0.1% soln. 0.1% oint.	1 mg	5 mg/day	5 mg/day
3. Prednisolone acetate	1% soln.	—	60-100 mg/day	—
4. Methyl prednisolone	—	20 mg/0.5 ml	—	30 mg/kg
5. Triamcinolone	0.1% oint.	20 mg/0.5 ml	—	—
6. Medrysone	1% susp.	—	—	—
7. Rimexolone	1% susp.	—	—	—
8. Fluorometholone	0.1% susp.	—	—	—

soln.: solution, oint.: ointment, susp.:suspension

Diclofenac sodium is a phenylacetic acid derivative and used in the management of ocular pain. The recommended dose is 50 mg twice daily.

The systemic administration of NSAIDs appears to have poor ocular penetration. NSAIDs must be administered with caution as they can cause gastric irritation, gastrointestinal bleeding, skin rashes and hypersensitivity reaction. They are contraindicated in acute peptic ulcer, bleeding disorders, aspirin induced allergy and asthma.

Topical NSAIDs

Topical NSAIDs are increasingly used in ophthalmic practice because of their safety, better penetration in the eye and effectiveness in a host of eye diseases. They inhibit prostaglandin release and act as a postoperative anti-inflammatory agent with analgesic properties. Preoperative use of topical NSAIDs not only inhibits intraoperative miosis but also reduces the risk of cystoid macular edema. They are indicated in phlyctenular conjunctivitis, vernal keratoconjunctivitis, episcleritis, scleritis, corneal limbal ulcers, postoperatively after an intraocular surgery, radial keratotomy and photorefractive keratectomy to reduce the pain.

Table 10.3: Topical NSAIDs

NSAIDs	*Concentration (%)*
1. Indomethacin	0.5-1
2. Diclofenac sodium	0.1
3. Flurbiprofen	0.03
4. Ketorolac tromethamine	0.5

Commonly available topical NSAIDs are listed in Table 10.3.

Topical NSAIDs must be used 3 to 4 times a day for 4 to 8 weeks in external ocular diseases. For inhibition of intraoperative miosis and prevention of postoperative CME, flurbiprofen (0.03%) or diclofenac sodium should be used 4 times daily 2 days before surgery, half-hourly for 2 hours before surgery and 4 times daily after surgery for 3 months. The main advantage of topical NSAIDs is their freedom from the side-effects of systemic NSAIDs. However, burning or stinging of the eyes and occasional photosensitivity or keratitis punctata may occur.

ANTIALLERGIC AGENTS

Mast-Cell Stabilizers

Mast-cell stabilizers block the calcium channel that is vital for cell degranulation thereby

stabilizing the cell. They inhibit the degranulation of both sensitized and nonsensitized mast-cells, thus, preventing the release of histamine and slow releasing substance of anaphylaxis (SRS-A).

Cromolyn sodium has no antihistaminic, sympathomimetic or corticosteroid-like action. The drug does not interfere with the antigen-antibody reaction but it suppresses the response to this reaction. Cromolyn sodium is topically used as 2 to 4 percent drops in the treatment of vernal keratoconjunctivitis, particularly in patients who are high responders to steroids. *Lodoxamide tromethamine* (0.1%) has cromolyn sodium -like action and it quickly relieves the symptoms of vernal keratoconjunctivitis. *Nedocromil sodium* (2%) is another mast-cell stabilizer used for treating allergic conjunctivitis.

Antihistamines

Topical antihistamines bind to H_1 receptors in the conjunctiva and reduce the itching. *Levocabastine* (0.05%) and *Emedastine* (0.05%) are commonly used topical ocular antihistaminics.

Multiple Action Agents

Multiple action agents have both mast-cell stabilizing and antihistamine properties. *Olopatadine* (0.1%), *Ketotifen fumarate* (0.025%), *Azelastine* (0.05%) and *Epinastine* (0.05%) are effective antiallergic eye drops with dual mode of action.

IMMUNOSUPPRESSIVE AGENTS

Immunosuppressive therapy is often recommended for the treatment of refractory uveitis, severe scleritis, ocular pemphigoid, high-risk keratoplasty and Graves ophthalmopathy. The therapy suppresses the clones of immunocompetent cells. Corticosteroids, cyclosporine, azathioprine, methotrexate, antilymphocyte serum and monoclonal antibodies are important immunosuppressive agents currently used in ophthalmic practice.

Cyclosporine A is an effective agent with a few systemic cytotoxic reactions. The mode of action of the drug is unknown. It is given orally in doses of 5 mg/kg daily (maintenance dose 1-3 mg/kg daily) in combination with corticosteroids. Cyclosporine A 1% drops are used topically in the treatment of refractory vernal keratoconjunctivitis and dry eye syndrome. Cyclosporine A, azathioprine, cyclophosphamide and chlorambucil are indicated for the treatment of sympathetic ophthalmia, Behçet's disease, Vogt-Koyanagi-Harada (VKH) syndrome, pars planitis and serpiginous choroiditis. However, cyclosporine is nephrotoxic and hepatotoxic and may cause hypertension and tremors.

Azathioprine, an antimetabolite, suppresses T-cells, however, it does not suppress humoral antibodies.

Methotrexate, also an antimetabolite, suppresses both humoral and cell-mediated immune reactions. It has anti-inflammatory action and diminishes chemotaxis.

DRUGS ACTING UPON INTRAOCULAR MUSCLES

Drugs are often used in ophthalmic practice for dilatation or constriction of the pupil. The pupil dilating drugs are known as *mydriatics* and pupil constricting, *miotics*. The drugs which are employed for paralyzing the accommodation or for paralyzing the ciliary muscle are called *cycloplegics*. In general, all mydriatics also paralyze the accommodation to a varying extent. Similarly, all miotics cause contraction of the ciliary muscle resulting in a state of partial or complete accommodation. All these drugs when instilled into the conjunctiva are absorbed through the cornea.

Mydriatic and Cycloplegic Drugs

These drugs can be divided into two groups: (i) parasympatholytic, and (ii) sympathomimetics.

Parasympatholytic Drugs

Atropine is a strong parasympatholytic mydriatic agent which causes paralysis of the sphincter pupillae and the ciliary muscle. It abolishes the action of acetylcholine (anticholinergic action) and, thus, causes mydriasis. It is used as 1% solution/ointment of atropine sulphate. It produces dilatation of pupil in about 45 minutes and paralysis of accommodation or cycloplegia in about two hours. The effect of the drug lasts for 7 to 10 days. Atropine is used for determination of refractive error in children and in adults with hypermetropia, relaxing the ciliary body in iridocyclitis and penalizing the better eye in amblyopia therapy. Fever and flushing of face may occur in children due to systemic absorption of atropine and contact dermatitis may be its local side effect.

Homatropine hydrobromide 2% is a synthetic compound that causes rapid mydriasis but incomplete cycloplegia, hence, may be employed for the determination of refraction. Its effect passes off entirely in 48 hours.

Tropicamide (0.5-1%) causes mydriasis in 20 minutes. It is the shortest acting mydiatric, the effect lasts for approximately 4-6 hours.

Cyclopentolate (1%) produces mydriasis in 30 minutes. It has more cycloplegic action than mydriatic action. Its effect lasts for approximately 12 to 24 hours.

Sympathomimetic Drugs

Adrenaline (Epinephrine) 1 in 1000 (1 mg/ml) to 1 in 10000 (0.1 mg/ml) acts directly on the dilator pupillae and causes dilatation of the pupil when used in irrigating solution. Preservative-free preparation is used intracamerally during an intraocular surgery.

Phenylephrine hydrochloride 5-10% induces mydriasis and vasoconstriction. It acts rapidly to produce mydriasis with minium cycloplegia in 30 minutes after instillation. A concentration of 2.5% is recommended for safe use in infants and pediatric patients.

Cocaine hydrochloride 2-10% is a local anesthetic which stimulates the sympathetic nerve endings in the dilator pupillae and causes moderate dilatation of the pupil.

Miotic Drugs

Parasympathomimetic Miotics

Acetylcholine chloride 1:100 directly acts on acetylcholine receptors of sphincter pupillae to induce miosis. It has extremely short duration of action, hence used as intraoperative miotic agent in cataract surgery.

Pilocarpine (0.5-6%) causes constriction of the pupil by directly stimulating the myoneural junctions of the sphincter muscle. It also produces contraction of the ciliary muscle. Its action is not longlasting, therefore has to be instilled 6-8 hourly.

Carbachol (Carbamyl-choline chloride) 0.01% is a short acting miotic used intracamerally during an intraocular surgery. It stimulates the motor endplate and inhibits acetylcholinesterase.

ANTIGLAUCOMA DRUGS

Antiglaucoma drugs or ocular hypotensive agents are described in the chapter of *Glaucoma*.

Antimetabolites

5-Fluorouracil (5-FU), a pyrimidine analogue, is used as an adjunct in trabeculectomy when there is a risk of failure of surgery. 5-FU is an antimitotic agent that inhibits fibroblastic proliferation and

prevents excessive postoperative scarring. 0.1 ml injection containing 5 mg of 5-FU is given daily or on alternate day basis through subconjunctival route upto a total dose of 50 mg. Corneal epithelial erosion and wound leak are common complications of this antifibrosis agent.

Mitomycin C (MMC) is isolated from *Streptomyces caespitosus*. It is an alkylating agent that inhibits DNA synthesis. The mode of action of MMC mimics that of ionizing radiation, therefore, it is also known as '*radiomimetic*'. A sponge soaked in 0.2-0.4 mg/ml of MMC is applied subconjunctivally on the scleral bed for 1-3 minutes during glaucoma or pterygium surgery. Mitomycin-augmented surgery prevents excessive postoperative scarring and, hence, reduces the risk of failure of filtering surgery or recurrence of pterygium. Common complications because of MMC use in glaucoma surgery are cataract formation, bleb infection and endophthalmitis while in pterygium excision they include scleral thinning and cataract. Postoperatively MMC, 0.2-0.4 mg/ml, topical preparation can be advocated after pterygium surgery instead of its intraoperative application.

VISCOELASTIC SUBSTANCES

Viscous and viscoelastic substances (VES) have increasingly been used in ocular microsurgery. Ideally, a viscoelastic substance should be inert, crystal clear, hydrophilic, elastic and viscous. Its viscosity creates space (deep anterior chamber or capsular bag distention) even under positive pressure and facilitates intraocular maneuvers safely. The coating property of the substance protects the corneal endothelium, iris, lens capsule and anterior hyaloid from the instruments and intraocular lens (IOL) intraoperatively.

The viscoelastic substances can either be cohesive or dispersive. *Cohesive VES* have high viscosity and are best at creating and preserving spaces. *Dispersive VES* are low viscosity agents that coat the ocular tissues and protect them from surgical trauma. Several preparations of viscoelastic substances are available. Some of the common ones are described below.

1. *Methylcellulose (Hydroxypropyl methylcellulose 2%)* is mainly viscous and barely elastic.
2. *Hypromellose (2%)* is like methylcellulose.
3. *Sodium hyaluronate (1%)* is a highly viscous and elastic substance. It is a cohesive VES that provides superb space maintenance. It protects the ocular tissues far better from the possible trauma from unfolding of IOL during implantation of foldable lenses than methylcellulose.
4. *Chondroitin sulfate* is a natural compound of connective tissue and is less elastic than sodium hyaluronate.
5. A combination of 4% chondroitin sulfate with 3% sodium hyaluronate is a dispersive VES that provides excellent tissue protection from intraoperative manipulations and reasonably good space maintenance.

The viscoelastic substances are used for following purposes:

1. IOL implantation
2. Phacoemulsification: The viscoelastic substances are used to protect the corneal endothelium, create more space by deepening the anterior chamber, dilate a poorly dilating pupil, tear the lens capsule during capsulorhexis and push the iris back in case of positive vitreous thrust during phacoemulsification.
3. Corneal transplantation
4. Reconstruction of the anterior segment following trauma
5. Removal of intraocular foreign body.

The use of viscoelastic substances is not totally free from side effects, postoperative transient rise in intraocular pressure is frequently encountered. Therefore, removal of viscoelastic material after completion of surgery through irrigation-aspiration is recommended. Considering the cost involvement and side effects, some eye surgeons

Table 10.4: Ocular toxicity of systemic drugs

Name of the drug	*Symptom*	*Sign*
1. Antitubercular		
a. Ethambutol	Photophobia, central scotoma, visual loss	Visual field defects, optic neuritis, optic atrophy, retinal hemorrhage
b. Isoniazid	Visual impairment, scotoma	Papilledema, optic neuritis, optic atrophy
c. Streptomycin	Scotoma, loss of vision	Nystagmus, optic neuritis, optic atrophy
2. Antiparasitic		
a. Quinine	Disturbance in vision especially for near, scotoma	Peripheral contraction of visual field, accommodation deficiency, cherry-red spot at macula, toxic amblyopia
b. Chloroquine	Visual impairment, swelling of the eye, central scotoma	Swelling of the conjunctiva, deposits in the corneal epithelium, bull's eye maculopathy
3. Antiemetic Chlorpromazine	Visual disturbance, scotoma, difficulty in near work	Weakness of accommodation, spasmodic upward deviations of eyes, deposits in corneal and lens epithelium, bull's eye maculopathy
4. Barbiturates	Headache, visual impairment	Nystagmus, poor convergence, ptosis, miosis/mydriasis, cortical blindness
5. Cardiac glycosides	Photophobia, xanthopsia, scotoma, poor night vision, defective color vision, diplopia, hallucination	Low intraocular pressure, optic neuritis, cortical blindness
6. Nonsteroidal anti-inflammatory drugs		
a. Aspirin	Scintillating scotoma, dryness of eyes	Nystagmus, hyphema, keratitis, mydriasis, papilledema, toxic amblyopia
b. Indomethacin	Poor night vision, color vision defect, diplopia	Retinal pigmentation, retinal edema, papilledema, toxic amblyopia
c. Phenylbutazone	Redness of eyes, dryness of eye, color vision defects	Conjunctival injection, corneal vascularization, retinal hemorrhages, toxic amblyopia
d. Ibuprofen	Swelling of lids	Lid edema, optic neuritis, toxic amblyopia
7. Oral contraceptives	Visual disturbances, scintillating scotoma, diplopia, contact lens intolerance	Myopia, nystagmus, corneal edema, occlusion of central retinal vein and artery, papilledema (intracranial hypertension)
8. Vitamins		
a. A	Diplopia, blurred vision	Nystagmus, exophthalmos, retinal hemorrhages, papilledema (intracranial hypertension)

Contd...

Table 10.4 contd...

Name of the drug	Symptom	Sign
b. D	Discomfort in the eyes	Calcium deposits in conjunctiva, cornea and sclera, band keratopathy, strabismus
9. Ergot	Scintillating scotoma	Nystagmus, retinal edema
10. Antineoplastic		
a. Adriamycin	Lacrimation	Conjunctivitis
b. Busulfan	Dryness of eyes	Cataract
c. Methotrexate	Photophobia, watering, irritation	Cataract
d. Tamoxifen	Foggy vision	Corneal opacity, maculopathy, intraretinal lipid deposits
e. Vincristine	Photophobia, diplopia	Corneal hyperesthesia, ptosis

prefer air or balanced salt solution (BSS) as alternatives to viscoelastic substances.

OCULAR SIDE EFFECTS OF SYSTEMIC DRUGS

Adverse effects of topical and systemic drugs used in ophthalmic practice have been described under the heading of individual drug. Some of the systemic drugs when administered for the treatment of extraocular disorders cause adverse ocular effects. A few drugs are neurotoxic and may lead to irretrievable blindness. Therefore, a general physician should know the ocular toxicity of commonly prescribed drugs. Important side effects of commonly used systemic drugs are listed in the Table 10.4.

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CHAPTER

11 Diseases of the Conjunctiva

ANATOMY

The conjunctiva is a translucent membrane which covers the posterior surface of the lids and then reflected onto the anterior part of the eyeball upto the margin of the cornea (limbus). It has 3 parts: the *palpebral conjunctiva* lining the eyelid, the *bulbar conjunctiva* covering a part of the eyeball and the *fornix* which unites the two (Fig. 11.1).

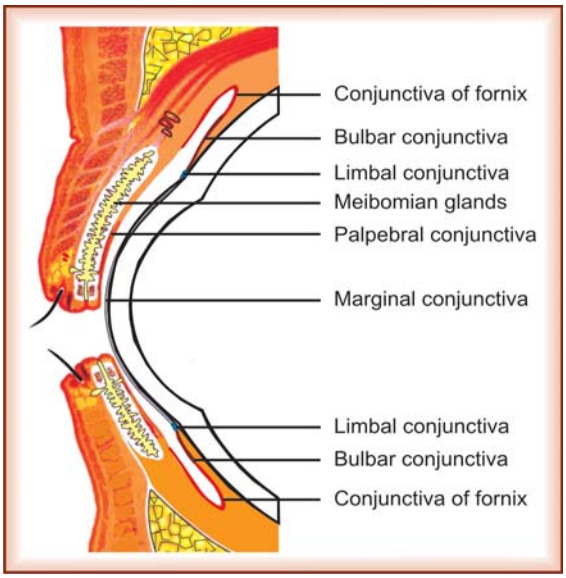

Fig. 11.1: Parts of conjunctiva

The *palpebral conjunctiva* is divided into marginal, tarsal and orbital zones. The *marginal conjunctiva* forms a transitional zone between the skin of the lid and the conjunctiva proper. It is continuous for about 2 mm on the back of the lid forming the subtarsal fold. The *tarsal conjunctiva* is firmly adherent to the tarsus of the upper lid, while in the lower lid it is only adherent to the breadth of the tarsus. It is highly vascular. The tarsal glands shine through it as yellow streaks. The *orbital* part of the conjunctiva lies loosely between the upper border of the tarsal plate and the fornix.

The *bulbar conjunctiva* covers the anterior part of the sclera. It is freely movable over the sclera excepting a zone of 3 mm width around the cornea (*limbal conjunctiva)* and at the insertions of the rectus muscle tendons. The *limbus* is a circular transitional zone between the cornea on one hand and the conjunctiva and the sclera on the other. The epithelium here is several layers thick and irregularly arranged. It shows papilliform digitations and contains blood vessels, lymphatics and melanin pigments.

The *forniceal conjunctiva* is a continuous *cul-de-sac* which is interrupted on the medial side by the caruncle and plica. It may be divided into a superior, an inferior and a lateral fornix.

Histologically, the conjunctiva consists of 3 layers: *epithelial*, *adenoid* and *fibrous*. There are two layers of epithelium over the palpebral conjunctiva. The layers gradually increase in number from the fornix to the limbus. The adenoid layer consists of loose connective tissue containing lymphocytes, mast cells and histiocytes. The adenoid layer does not develop until after the first 2 or 3 months of life, hence, follicles do not appear in early infancy. The fibrous layer is a thick meshwork of collagen and elastic fibres which blends with Tenon's capsule.

Goblet cells, serous glands and accessory serous glands are found in the conjunctiva. Numerous mucus secreting goblet cells are present in the epithelium of bulbar conjunctiva and fornix. They are true unicellular mucous glands which moisten the conjunctiva and the cornea by discharging mucin.

Plica semilunaris is a vestigeal structure in human beings. It represents the third eyelid or the nictitating membrane of lower vertebrates. It is a crescent-shaped fold of conjunctiva found at the inner canthus with its concavity towards the cornea.

Caruncle is a small fleshy ovoid body measuring 5 mm × 3 mm situated in the lacus lacrimalis to the medial side of plica semilunaris. It is covered by stratified squamous epithelium and contains hair follicles and sebaceous and sweat glands.

Blood Supply of the Conjunctiva

The conjunctiva derives its blood supply from:

1. Palpebral branches of nasal and lacrimal arteries of the lids and
2. Anterior conjunctival arteries, the branches of anterior ciliary arteries.

The palpebral conjunctiva is supplied by the post-tarsal plexus of the upper and lower lids. The perforating branches from the peripheral palpebral arcade supply the fornix, their descending branches anastomose with the marginal arcade and ascending branches continue in the bulbar conjunctiva as the posterior conjunctival artery and supply the whole of the bulbar conjunctiva excepting a zone 4 mm wide around the limbus. The terminal branches of the posterior conjunctival artery anastomose freely with the anterior conjunctival artery forming a pericorneal plexus.

The conjunctival veins drain either in the post-tarsal venous plexus of the lid or in the superior or inferior ophthalmic vein.

Lymphatics of conjunctiva lie superficially as well as deep and form an irregular network. Lymphatics of the palpebral conjunctiva join the lymphatics of lids. The lymph vessels from the lateral side drain into the preauricular lymph nodes and those from the medial side into the submandibular nodes.

Nerve Supply of the Conjunctiva

The sensory nerve supply of the conjunctiva is derived from the trigeminal nerve—from the infratrochlear branch of nasociliary nerve, supratrochlear and supraorbital branches from the frontal nerve, the lacrimal nerve and the infraorbital nerve. The ciliary nerves supply the limbal conjunctiva. The sympathetic nerves come from the sympathetic plexus along the branches of the ophthalmic artery.

Bacterial Flora of the Conjunctiva

The conjunctiva is practically never free from organisms. The eyes of infants harbor a number of bacterial species including *S. aureus*, *S. epidermidis*, *Streptococci* and *E. coli*. With increasing age, gram-negative bacteria invade the conjunctiva. *Propionibacterium acnes* and *Corynebacterium xerosis* can be isolated from the healthy conjunctiva.

A relatively low temperature of the conjunctiva due to constant evaporation of tears, mecha-

nical action of the lids, pumping action on the tear drainage system, constant epithelial exfoliation and a moderate blood supply make the conjunctiva unsuitable for the propagation of organisms. Further, tears contain lysozymes, betalysins, IgA and IgG, all of which inhibit bacterial growth. Nevertheless, the conjunctiva is quite frequently implicated in diseases because it is exposed to all types of exogenous irritants and infections. Moreover, it is prone to allergic reactions and often gets involved in metabolic disorders.

DISEASES OF THE CONJUNCTIVA

The diseases of the conjunctiva may be described under following heads:

1. Symptomatic conditions of the conjunctiva
2. Inflammation of the conjunctiva (conjunctivitis)
3. Degenerations of the conjunctiva
4. Cysts and tumors of the conjunctiva.

Symptomatic Conditions of the Conjunctiva

Symptomatic conditions include hyperemia, chemosis, ecchymosis, xerosis and pigmentation of the conjunctiva.

Hyperemia of the Conjunctiva

Hyperemia or congestion of the conjunctival vessels may be transient or chronic.

Etiology The transient hyperemia is due to irritation by a foreign body (eyelash, coal particle, concretion, etc.). The removal of the irritant provides prompt relief.

Adverse atmospheric conditions, especially dry dusty climate, refractive errors, metabolic disorders such as gout and diabetes, allergic predispositions, over-indulgence in smoking and alcohol, insomnia and exposure to strong light, may cause recurrent or chronic hyperemia of the conjunctiva.

Clinical features The patient complains of discomfort in the eye often associated with grittiness, heaviness and tiredness. The eye appears normal except for mild to moderate congestion towards the fornices.

Treatment Symptomatic relief may be obtained by instillation of an astringent lotion like zinc sulphate (0.25%) or a decongestant eye drop like levocabastine and naphazoline, but for lasting cure the primary factor causing hyperemia should be removed.

Chemosis of the Conjunctiva

Chemosis or edema of the conjunctiva is quite common.

Etiology Chemosis occurs due to laxity of the tissue and seen in ocular and systemic diseases.

The local causes of chemosis of conjunctiva include acute conjunctivitis, keratitis, corneal ulcer, iridocyclitis, orbital cellulitis, panophthalmitis and acute congestive glaucoma. It may also be associated with orbital tumors and thyroid exophthalmos owing to venous stasis.

Systemic diseases such as nephritis, congestive heart failure, hypoproteinemia and allergic reactions (drug allergy, urticaria, angioneurotic edema) frequently produce chemosis of the conjunctiva.

Clinical features The conjunctiva becomes swollen and appears gelatinous because of exudation from the capillaries. The collection of exudate is most prominent in bulbar and forniceal conjunctiva.

Treatment The management of chemosis includes treatment of the underlying cause.

Ecchymosis of the Conjunctiva

Ecchymosis or subconjunctival hemorrhage is often seen in children and aged people.

Etiology Ecchymosis is found in acute conjunctivitis, especially in acute hemorrhagic conjunctivitis and conjunctivitis caused by *Streptococcus pneumoniae* and *Haemophilus aegyptius* (Koch-Weeks bacillus). Trivial trauma causes rupture of the conjunctival capillaries leading to small subconjunctival hemorrhage, while fracture of the base of skull or a violent whooping cough gives rise to large subconjunctival hemorrhage. In fracture of the base of skull, the blood seeps along the floor of the orbit and appears under the conjunctiva within 12 to 24 hours after the injury.

Hemorrhages are also seen after crush injuries due to pressure on thorax and abdomen. Blood dyscrasias, scurvy, diabetes, arteriosclerosis and hypertension are the other important causes of ecchymosis.

Clinical features Most cases of subconjunctival hemorrhage are symptomless. However, ecchymosis due to conjunctivitis or trauma gives annoying symptoms. The hemorrhage may be petechial or an extensive one covering the bulbar conjunctiva (Fig. 11.2). The latter gives an alarming picture.

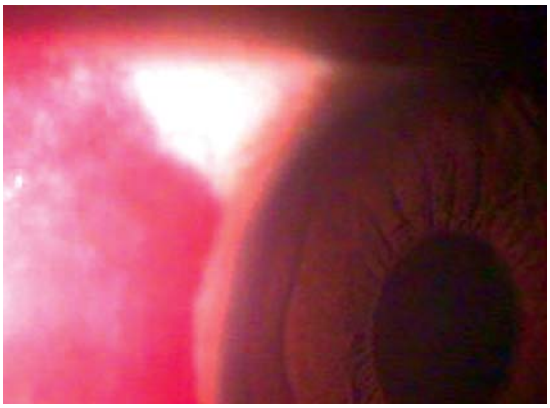

Fig. 11.2: Subconjunctival hemorrhage

Treatment Generally, the subconjunctival hemorrhage gets absorbed by itself within two to three weeks. Cold compresses check the bleeding in the initial stages. Most cases do not require any treatment except reassurance.

Xerosis

Xerosis is defined as a dry lusterless condition of the conjunctiva which manifests in two forms:

1. *Parenchymatous xerosis*: A sequel to local disease of the conjunctiva involving all its layers, and
2. *Epithelial xerosis*: Associated with vitamin A deficiency.

Parenchymatous xerosis Parenchymatous xerosis is a cicatricial degeneration of the conjunctiva following widespread destructive interstitial conjunctivitis as seen in trachoma, membranous conjunctivitis, pemphigus or pemphigoid conjunctivitis and physical/chemical burns. Severe degree of xerosis is seen in long-standing proptosis, ectropion and lagophthalmos following exposure.

Epithelial xerosis (Xerophthalmia) Xerophthalmia is a term applied to all ocular manifestations of impaired vitamin A metabolism from night-blindness to more or less complete corneal destruction. It is responsible for nearly 100000 new cases of blindness worldwide each year.

Etiology Xerophthalmia results either from an inadequate supply of vitamin A or a defective absorption from the gut due to gastrointestinal disorders. Epithelial xerosis is predominantly a disease of children under 5 years of age coming from lower socio-economic strata. They are usually ill-nourished, ill-looking and marasmic. Concurrent infections with measles, microbial agents and herpes simplex may predispose the child to keratomalacia.

Main pathological changes are found in the epithelium which assumes epidermoid character like skin (epidermidalization of the conjunctival epithelium) with granular and horny layers. Owing to the destruction of goblet cells, the mucus is not secreted and dry, dull or pigmented spots appear in the conjunctiva. Vicarious secretion from the meibomian glands is deposited on these spots, so the tear film fails to moisten them. *Corynebacterium xerosis* grows abundantly in xerotic conjunctiva.

Classification For diagnostic and therapeutic purposes, the following WHO classification of xerophthalmia is used:

XN	Night-blindness
X1A	Conjunctival xerosis
X1B	Bitot's spots
X2	Corneal xerosis
X3A	Corneal ulceration/keratomalacia affecting less than one-third corneal surface
X3B	Corneal ulceration/keratomalacia affecting more than one-third corneal surface
XS	Corneal scar due to xerophthalmia
XF	Xerophthalmic fundus.

Clinical features:

- *XN—Night-blindness* is the earliest symptom of xerophthalmia.
- *X1A—Conjunctival xerosis* is characterized by lack of luster of the conjunctiva associated with its wrinkling owing to the loss of elasticity. The wrinkling of the conjunctiva can be seen on lateral movements of the eye as the conjunctiva forms crescents along the limbus.
- *X1B—Bitot's spot* is a white, foamy lusterless triangular (base at the limbus and apex towards the outer canthus) plaque invariably situated on the bulbar conjunctiva (Figs 11.3 and 11.4). It is superficial and raised above the surface of the conjunctiva. It is usually bilateral and temporal, and less frequently nasal.

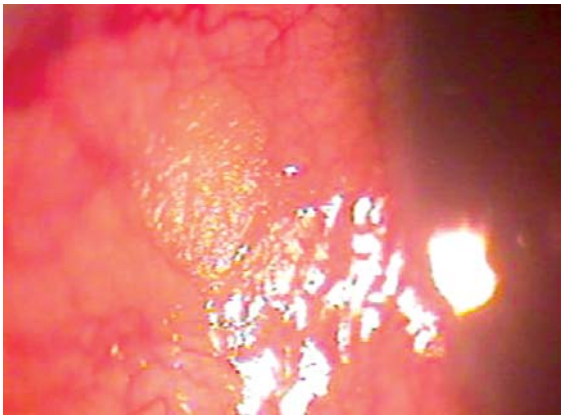

Fig. 11.3: Bitot's spot

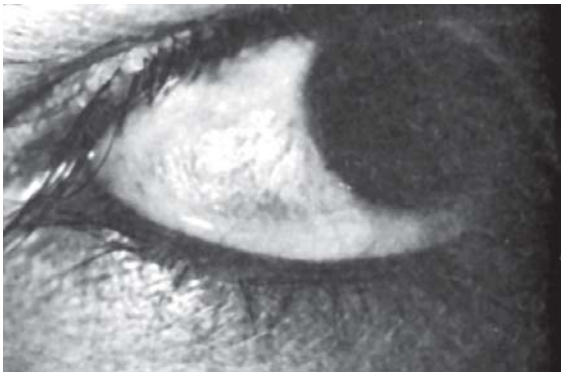

Fig. 11.4: Bitot's spot with keratinization

- *X2—Corneal xerosis* manifests into two forms: *precorneal xerosis* wherein there occurs loss of corneal luster and decreased corneal sensitivity and *true corneal xerosis* in which cornea lacks luster and its surface becomes pebbly. Sometimes keratinized plaques may be formed on the cornea.
- *X3A, X3B—Corneal ulceration/keratomalacia* is a late manifestation of xerophthalmia in which less than one-third of the corneal stroma melts away due to colliquative necrosis (X3A). In Keratomalacia (X3B) more than one-third of the cornea is involved. The cornea appears cloudy and soft. The sloughing of the necrotic stroma leaves a large ulcer which may perforate (Fig. 11.5).

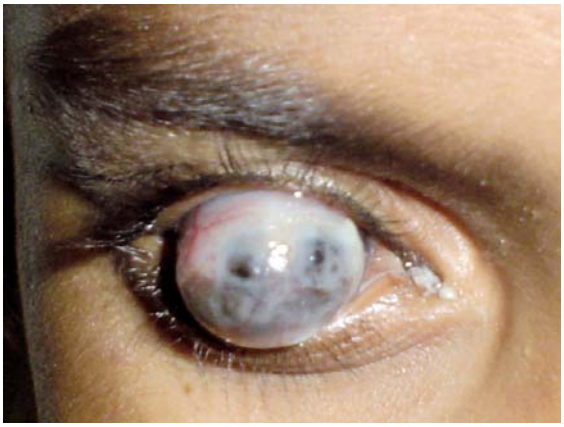

Fig. 11.5: Keratomalacia (*Courtesy*: Prof Manoj Shukla and Dr Prashant Shukla, AMUIO, Aligarh)

- *XS—Corneal scars* are of different densities and are left after healing of ulcers. If they cover the pupillary area, visual acuity is grossly impaired.
- XF—*Xerophthalmic fundus* lesions appear as small, discrete, yellow dots in the peripheral fundus. Perhaps, they represent a focal depigmentation of the retinal pigment epithelium.

Treatment

The parenchymatous xerosis is a preventable condition. Prompt treatment of trachoma or membranous conjunctivitis should be carried out. Adequate precautions should be taken to avoid ocular chemical burns. The tear substitutes and mucous grafting are often needed.

The epithelial xerosis in infants can be prevented by administering prophylactic vitamin A in mothers during pregnancy. Breastfeeding should be encouraged. Proper treatment of gastrointestinal disturbance, particularly worm infestations, is necessary. Methyl cellulose or lubricating eye drops are used locally. If secondary infection is feared, topical antibiotic is added. X3A and X3B cases need treatment on the lines of corneal ulcer. Generally, the daily requirement of vitamin A for a child is 3000 to 4000 IU. It should be supplemented with protein-rich diet to correct protein-energy-malnutrition (PEM) and to facilitate the absorption of vitamin A. In mild to moderate degree of xerophthalmia, dietetic correction with the inclusion of vitamin A rich green vegetables, carrot, butter, egg, fish, cod-liver or halibut-liver oil, gives satisfactory results. The WHO recommended a dose of 200000 IU of vitamin A in 3 doses for the management of clinical xerophthalmia (Table 11.1).

Conjunctival Pigmentation

The conjunctiva may show discoloration in following systemic and local conditions:

1. It becomes yellow in jaundice due to presence of bile pigments.
2. Brown to slaty discoloration of conjunctiva is found in Addison's disease or chronic adrenal insufficiency.

Table 11.1: WHO recommended vitamin A therapy for xerophthalmia

Patients	*Age*	*Dose*	*Schedule*
Children	< 12 months	100000 IU	1st day, 2nd day and repeat 2-4 weeks later
Children	12 months or older	200000 IU	1st day, 2nd day and repeat 2-4 weeks later
Women with NB or Bitot's spot	Child-bearing age	10000 IU 25000 IU	Daily for 2 weeks or Weekly for 4 weeks
Women with corneal lesions		200000 IU	1st day, 2nd day and repeat 2-4 weaks later

NB: Night-blindness

3. A characteristic symmetrical semilunar accumulation of brown or gray pigments in the sclera and/or bulbar conjunctiva is found in ochronosis wherein an incomplete metabolism of tyrosine (alkaptonuria) and phenylalanine occurs.
4. The conjunctiva becomes red in subconjunctival hemorrhage and later leaves a brown pigmentary spot.
5. Benign melanoma of the conjunctiva and precancerous melanosis of the conjunctiva impart brown-black pigmentation. Local application of soot *(Kajal)* or mascara (often used by females) leads to black pigmentation of conjunctiva.
6. Iatrogenic brownish staining of the conjunctiva is known as *argyrosis*. It was common due to prolonged application of silver salts for the management of trachoma in the past and resulted in impregnation of reduced metallic silver in the elastic tissue of the conjunctiva.
7. Long-term topical use of adrenaline in glaucoma patients may cause formation of black spots in the conjunctiva owing to oxidation of adrenaline to melanin.

INFLAMMATION OF THE CONJUNCTIVA (CONJUNCTIVITIS)

Conjunctivitis is the most common eye disease worldwide. It is usually of two types:

1. Infectious and
2. Noninfectious.

The noninfectious conjunctivitis may further be subdivided into:

a. Allergic
b. Toxic
c. Traumatic
d. Secondary, and
e. Idiopathic.

Infectious Conjunctivitis

A wide variety of etiological agents, bacteria, virus and fungi, can cause infection in the conjunctiva. There is no uniform criterion for the classification of infective conjunctivitis. Depending on the onset it may be divided into two broad clinical categories: *acute* and *chronic*.

The etiology of infective conjunctivitis has shown a remarkable change in the recent past. During preantibiotic era, bacterial conjunctivitis dominated. But after the middle of the twentieth century, 75% cases of conjunctivitis were found to be nonbacterial in origin in a survey conducted in London. Viruses were responsible for 35% of conjunctivitis. In the East, outbreaks of bacterial conjunctivitis still occur during each premonsoon period which may or may not be associated with rickettsial or viral conjunctivitis.

To facilitate description, acute conjunctivitis may further be classified as acute catarrhal or mucopurulent, purulent, membranous and hemorrhagic.

Acute Catarrhal or Mucopurulent Conjunctivitis

Acute catarrhal conjunctivitis is an acute infective type of conjunctivitis characterized by hyperemia of the bulbar conjunctiva and papillary hypertrophy of the palpebral conjunctiva associated with mucopurulent discharge. The condition is commonly seen in children. However, it may affect any age group. It has a short incubation period (24-48 hours).

Etiology The disease is caused by *Staphylococcus aureus* (coagulase-positive), *Koch-Weeks bacillus*, *Pneumococcus* and *Streptococcus*. It may also occur in association with acute infective eruptive fevers such as measles and scarlet fever.

Clinical features Acute mucopurulent conjunctivitis may manifest either in a mild or a severe form. The former gives minimum symptoms, but the presence of hyperemia of conjunctiva and tags of mucus at the canthi help in the diagnosis. Quite erroneously, it is called *cold in the eyes*.

The severe form reaches its peak in 3 to 4 days. Heaviness or discomfort in the eye, glueing of the eyelashes of the upper and lower lids, particularly after the night sleep, photophobia and colored halos are the common symptoms. The conjunctiva becomes fiery red with marked papillary hypertrophy of the palpebral conjunctiva (Fig. 11.6) and congestion of vessels towards the fornices. The lids are slightly edematous. The mucopurulent discharge is found in the fornices and on the margin of the lids matting the lashes. The accumulation of mucus over the cornea results in colored halos due to the prismatic effect.

Complications The condition is benign but if untreated passes into a chronic phase. Staphylococcal mucopurulent conjunctivitis may cause superficial corneal erosions, while pneumococcal conjunctivitis shows petechial hemorrhages on the bulbar conjunctiva.

Treatment The treatment of mucopurulent conjunctivitis is essentially based on two principles: frequent irrigation of the conjunctival *cul-de-sac* to remove the discharge and control of the infection. The infected eye is washed 4 to 5 times a day with normal saline warmed at room temperature. The irrigation not only removes the mucus but dilutes the toxins and increases the flow of antibodies.

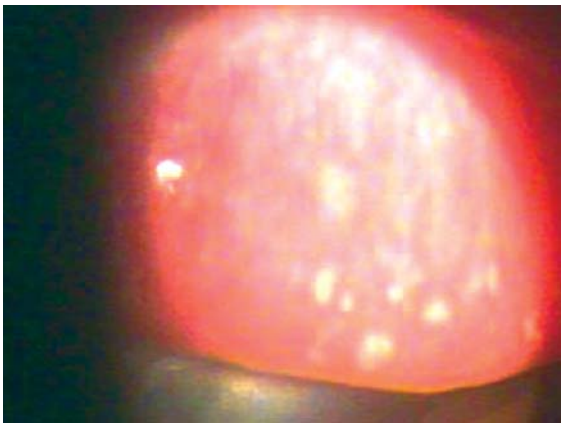

Fig. 11.6: Acute mucopurulent conjunctivitis

Ideally, the selection of an antibiotic or chemotherapeutic agent for the control of infection should be done after sensitivity test. However, it is not possible in practice. Therefore, one of the broad-spectrum antibiotics like ciprofloxacin 0.3%, ofloxacin 0.3%, gatifloxacin 0.3%, moxifloxacin 0.5% or chloramphenicol 0.5% is commonly used. An antibiotic ointment (ciprofloxacin, gatifloxacin, tetracycline or oxytetracycline) is applied at bed time to prevent the lids from sticking together. Dark glasses may be worn to minimize photophobia, but the eye should never be bandaged as this promotes the growth of organisms and enhances the accumulation of discharge.

Considering the contagious nature of the disease, prophylactic measures must be taken to check its spread in the family and community.

Acute Purulent Conjunctivitis

Acute purulent conjunctivitis is also known as *acute blenorrhea* and is marked by a profuse purulent discharge. The disease was rampant in the Middle East in the early part of the 20th century and caused untold miseries by its blinding sequelae. It occurs in two forms:

1. Purulent conjunctivitis of newborn (ophthalmia neonatorum), and
2. Purulent conjunctivitis of adult.

Purulent Conjunctivitis of Newborn (Ophthalmia Neonatorum)

Ophthalmia neonatorum is a bilateral conjunctivitis of newborn, characterized by copious purulent discharge, marked chemosis of the conjunctiva and swelling of the lids.

Etiology The disease is contacted during birth from the mother's infected genitourinary tract or from infected linen and fingers. A number of organisms, viz. *Neisseria gonorrhoeae*, *Staphylococcus aureus*, *Streptococcus pneumoniae*, *Staphylococcus*

hemolyticus and *E.coli* are established causative pathogens. Gonococcal ophthalmia neonatorum is a serious and violent condition, while *Chlamydia* and adenoviruses cause mild purulent conjunctivitis.

Causes of neonatal conjunctivitis can be separated on the basis of duration of onset of disease. The chemical conjunctivitis starts within a few hours after the application of silver nitrate drops (used for prophylaxis of ophthalmia neonatorum), gonococcal and meningococcal conjunctivitis 3 days after exposure and neonatal inclusion conjunctivitis and herpes simplex conjunctivitis 5 or more days after exposure (Table 11.2).

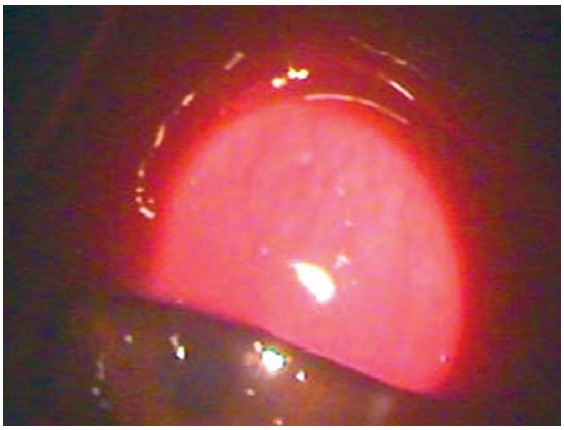

Fig. 11.7: Acute purulent conjunctivitis

Clinical features Ophthalmia neonatorum usually manifests in the first week after birth. Initially, a watery secretion is noticed from the baby's eye (normally tears are not secreted in the first six weeks of life, therefore, any secretion from the eye should be considered abnormal). It soon becomes mucopurulent and ultimately purulent. Both eyes are almost always involved. The infant is irritable and his conjunctiva intensely inflamed, chemotic and red (Fig. 11.7). The chemosis is so marked that the bulbar conjunctiva bulges through the lids and cornea appears to be situated at the bottom of a crater-like pit. The lids are swollen and brawny. The flakes of thick purulent discharge are seen over the conjunctiva and the lid margin. Both gram and Giemsa stains of the conjunctival scrapings help to identify *N. gonorrhea, C. trachomatis* and other causative organisms.

The disease has a short incubation period (1-3 days). If untreated, the acute phase lasts for 10-15 days and then the discharge diminishes and swelling gradually subsides.

Complications In gonococcal ophthalmia neonatorum, the corneal complication is a rule. The

Table 11.2: Diagnostic features of neonatal conjunctivitis

Causes	*Onset*	*Discharge*	*Smear and culture*
Silver nitrate (Crede's prophylaxis)	Within few hours	Slight watery or mucus	Negative culture
Gonococcal conjunctivitis	2-4 days	Copious purulent discharge	Intracellular gram-negative diplococci, culture positive on blood agar
Nongonococcal bacterial (*S. aureus, Streptococcus pneumoniae*)	4-5 days	Mucopurulent	Gram-positive or gram-negative organisms in smear and positive culture
Chlamydia (TR-IC infection)	5-14 days	Mucopurulent, occasionally purulent	Cytoplasmic inclusion bodies, negative culture
Herpes simplex infection	5-7 days	Watery	Multinucleated giant cells, cytoplasmic inclusion bodies and negative culture

organism is capable of invading the intact corneal epithelium; the corneal ulceration develops over an area just below the center of the pupil corresponding to the lower lid margin. The ulcer is prone to perforation. A mild to severe degree of iridocyclitis accompanies the ulcer. The perforation of ulcer gives many blinding sequelae, such as, leukoma adherence, partial or total anterior staphyloma, nystagmus and phthisis bulbi.

Treatment Ophthalmia neonatorum is a preventable disease. The prenatal diagnosis and treatment of birth canal infection should be carried out adequately. Aseptic measures must be taken at the time of delivery. Soon after birth, the lids of the infant be thoroughly cleaned with a piece of sterile gauze. Prophylactic medication either by adopting *Crede's method* or other regimen should be carried out. In Crede's method a drop of 1% silver nitrate is instilled in each eye of the infant soon after birth. The procedure may cause a mild chemical conjunctivitis which is self-limiting. Topical instillation of a combination of bacitracin and polymyxin B may also be used. Povidone-iodine 5% is commonly used as a prophylactic eye drop that does not cause any toxic reaction.

The infants with ophthalmia neonatorum require prompt treatment. The eye must be irrigated with warm saline at least four times a day. *Neisseria gonorrhoeae* infection is usually controlled by intensive antibiotic therapy. Earlier the standard regimen was instillation of penicillin drops, in a concentration of 5000 to 10000 unit/ml, every minute for half an hour, every five minutes for another half an hour and then half-hourly instillations till the infection is controlled. Owing to increasing prevalence of resistance to penicillin, topical therapy with tetracycline, gentamicin, bacitracin and fluoroquinolone is recommended. Topical ciprofloxacin 0.3% drops hourly and cephtriaxone 25 to 50 mg/kg IV or IM single dose or cephotaxime 25 mg/kg IM or IV 12 hourly are found to be very effective. Chlamydial infection can be controlled by topical erythromycin or tetracycline. Systemic erythromycin 12.5 mg per/kg oral or IV for 14 days is recommended to control mixed infection. Great care is needed to examine and treat the eye if the cornea is involved. Topical atropine eye ointment must be used but the eye must not be bandaged.

Acute Purulent Conjunctivitis of Adults

Acute purulent conjunctivitis of adults is often unilateral and associated with urethritis and arthritis.

Etiology The disease is venereal in origin and the infection is transmitted from genitals to the eye. Males are predominantly affected. The disease has a short incubation period. It is commonly due to *N. gonorrhoeae* but other organisms responsible for ophthalmia neonatorum can also cause the disease.

Clinical features Gritty sensation, photophobia, blurring of vision, pain in the eye and mild constitutional disturbances are common symptoms of the disease. The patient is generally in agony and does not allow ocular examination easily. There occurs brawny edema of the upper lid. The eyelashes are matted with organized thick discharge. The conjunctiva is markedly edematous and velvety in appearance.

Complications The cornea becomes hazy with central gray area of necrosis. Marginal ulcers usually develop due to retention of pus in the ballooned conjunctiva. Iridocyclitis may ensue even before perforation of the corneal ulcer. The patient is febrile and has enlarged and painful preauricular lymph nodes. In gonococcal conjunctivitis, urethritis is almost an invariable accompaniment. Arthritis, endocarditis and septicemia may also be found.

Treatment The basic principle of treatment of acute purulent conjunctivitis of adults is to protect the unaffected eye and a prompt control of

infection in the affected one. The other eye can be protected by using an eye shield. However, the most effective method is to institute prophylactic treatment in the healthy eye.

Repeated irrigation and intensive therapy with ciprofloxacin (0.3%) eye drop 2 hourly and erythromycin 1% eye ointment often bring improvement in the clinical picture. *Gonococcus* may be present in the conjunctiva for a long period, hence, the therapy should be continued for two to three weeks. Atropine is applied if the cornea and the uvea are involved. Analgesics are helpful in ameliorating the general symptoms.

The treatment of gonococcal conjunctivitis without septicemia in adults is a single dose of ceftriaxone 1g IM. However, patients with keratoconjunctivitis or disseminated gonococcal infection should be treated with ceftriaxone 1 g IV or IM 12 hourly for at least 3 days. Patients allergic to penicillin should be treated with spectinomycin 2 g IM as a one time dose or 12 hourly in divided doses. Oral ciprofloxacin and norfloxacin are also effective as well as economical.

Acute Membranous Conjunctivitis

Acute inflammation of the conjunctiva associated with the formation of a membrane or pseudomembrane on the palpebral conjunctiva (Fig. 11.8) characterizes acute membranous conjunctivitis.

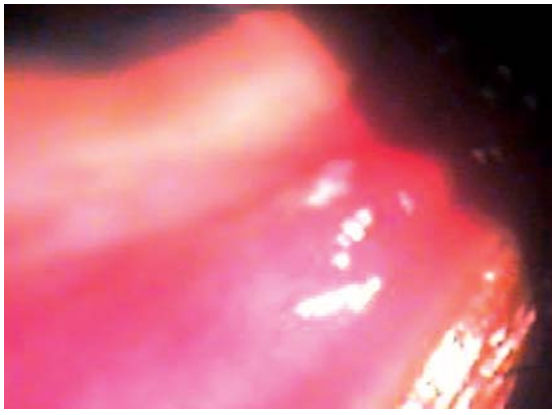

Fig. 11.8: Membranous conjunctivitis

Etiology The membranous conjunctivitis is more or less synonymous with diphtheritic conjunctivitis since *Corynebacterium diphtheriae* causes membrane formation. However, *Streptococcus hemolyticus, Streptococcus pneumoniae, Neisseria gonorrhoeae, Staphylococcus aureus, H. aegyptius, E. coli*, adenoviruses and herpes simplex virus can also produce membranous conjunctivitis. Erythema multiforme and alkali burn may also lead to membrane formation.

The membrane may be false (pseudo) or true, it appears as a result of coagulative response to infectious or toxic agents. In pseudomembrane a coagulum consisting of fibrin, mucus and pus is deposited on the surface of the epithelium, while in a true membrane the epithelial layers undergo coagulative necrosis. The removal of a pseudomembrane leaves an intact epithelium, while a raw bleeding surface is left behind following the removal of a true membrane.

Membranous conjunctivitis usually occurs in children between 2 and 8 years of age, who are not immunized. The disease may appear either in a mild or a severe form. Membranous conjunctivitis of diphtheritic origin is often severe. It is, however, sometimes seen that mild cases of membranous conjunctivitis may be diphtheritic and severe nondiphtheritic, especially streptococcal.Therefore, a confirmed diagnosis can be made only after the bacteriological examination.

Clinical features Mucopurulent discharge, mild degree of swelling of the conjunctiva and lids, a white pseudomembrane on the palpebral conjunctiva and regional lymphadenopathy may be seen in the mild variety of conjunctivitis.

In severe cases, the patient is toxic and acutely ill. Pain is often severe. The lids are swollen, red and tense making their eversion difficult.

The course of membranous conjunctivitis can be divided into 3 stages.

Stage of infiltration: The conjunctiva is markedly chemosed and infiltrated with semisolid exudates

which impair ocular motility and threaten the corneal transparency. An extensive true membrane is found to cover the entire palpebral conjunctiva; it is seldom found on the bulbar conjunctiva. The regional lymph nodes are usually enlarged and may undergo suppuration. The membrane may also be seen covering the throat or nasal mucosa in diphtheritic conjunctivitis.

S*tage of suppuration*: The acute phase lasts for 6 to 10 days during which cornea may ulcerate. Gradually, the necrosed conjunctiva sloughs out and appears red and succulent.

S*tage of cicatrization*: Adhesions (symblepharon) usually develop between the raw areas on the palpebral and the bulbar conjunctiva. The cicatrization of conjunctiva may lead to xerosis and entropion.

Treatment Proper immunization in infancy and quick isolation of the infected patient are the usual preventive measures. To start with, every case of membranous conjunctivitis must be treated as diphtherial unless proved otherwise by bacteriological examination. Immediate local and general treatment with penicillin is instituted. Antidiphtheritic serum (ADS) and penicillin drops (10000 unit per ml) are instilled hourly into the conjunctival sac. Atropine sulphate (1%) should be applied if cornea is involved. Intramuscular injections of antidiphtheritic serum (10000 unit) and crystalline penicillin (5 lacs unit) are given 12 hourly. Diphtheritic antitoxins given locally and systemically are effective when administered with antibiotic.

Use of contact shell may prevent symblepharon formation. Some cases may need plastic surgery with amniotic membrane transplantation.

For the management of nondiphtheritic conjunctivitis, treatment with topical and systemic antibiotic (depending on the sensitivity of the organism) is energetically instituted.

Herpes Simplex Virus Conjunctivitis

Acute conjunctivitis may also be caused by herpes simplex virus (HSV) type 1 and 2. Herpes simplex virus type 1 causes an acute unilateral blepharoconjunctivitis with vesicular lesions on the lids, intense papillary hypertrophy of the conjunctiva and classical dendritic lesion on the cornea. There occurs marked enlargement of the preauricular lymph glands. The virus can also produce a follicular conjunctivitis.

Herpes simplex virus type 2 conjunctivitis is essentially a venereal infection acquired by direct contamination of eye from birth canal. Primary HSV conjunctivitis is a self-limiting disease. Topical antiviral therapy with acyclovir 3% eye ointment controls the infection.

Acute Adenovirus Conjunctivitis

Adenoviruses are known to produce acute follicular conjunctivitis as seen in pharyngoconjunctival fever (PCF) and epidemic keratoconjunctivitis (EKC).

Pharyngoconjunctival Fever

Pharyngoconjunctival fever primarily affects children and appears in epidemic form. It is due to adenovirus serotypes 3, 4 and 7. Acute follicular conjunctivitis, pharyngitis, fever and preauricular lymphadenopathy are the characteristic signs. Systemic signs mimic influenza. Punctate keratitis may be the only corneal sign of the disease.

The conjunctivitis is self-limiting and there is no specific treatment but topical antibiotics should be used to control secondary bacterial infection.

Epidemic Keratoconjunctivitis

As is evident by the name, the keratoconjunctivitis occurs in widespread epidemics that mostly spreads through infected ophthalmic instruments especially tonometers.

Etiology Epidemic keratoconjunctivitis is caused by adenovirus serotypes 3, 7, 8 and 19. The definitive diagnosis is made after recovering the virus from eye and growing it in cell culture.

Clinical features EKC is characterized by photophobia, acute follicular or membranous conjunctivitis, subepithelial infiltrates in the cornea, scanty discharge and preauricular lymphadenopathy. Pseudomembrane on the palpebral conjunctiva develops predominantly. Petechial hemorrhages on bulbar conjunctiva and subconjunctival hemorrhages can occur.

Diffuse punctate epithelial keratitis is the earliest corneal lesion. Stromal corneal infiltrates develop within two weeks' time due to immune response to the adenovirus. Later, discrete anterior stromal infiltrates covering the pupillary area (Fig. 11.9) may appear which may persist for months or years causing visual disturbances.

Prophylaxis In order to prevent the spread of epidemic, cleaning and sterilization of all instruments that touch the patient's eye must be done.

Treatment The treatment of EKC is nonspecific and symptomatic. Broad-spectrum antibiotics are often used to prevent secondary infections. Topical corticosteroids are recommended in patients with conjunctival membrane or photophobia.

Newcastle Conjunctivitis

Newcastle conjunctivitis is a rare disorder occurring in small epidemics among poultry workers and is caused by Newcastle virus. The conjunctivitis is indistinguishable from pharyngoconjunctival fever.

Acute Hemorrhagic Conjunctivitis

An epidemic of acute hemorrhagic conjunctivitis occurred at the time when Apollo spacecraft was launched, hence, it is also known as *Apollo conjunctivitis*.

Etiology The etiological agents of acute hemorrhagic conjunctivitis are identified as coxsackie virus and enterovirus 70 belonging to picornavirus group. The disease affects all age groups but is mostly seen in young patients. It is contagious and its transmission appears to be by hand-to-eye contact.

Clinical features A sudden onset of mixed papillary and follicular hyperplasia, petechial and coalesced hemorrhages in the bulbar (Fig. 11.10) and the tarsal conjunctiva and preauricular

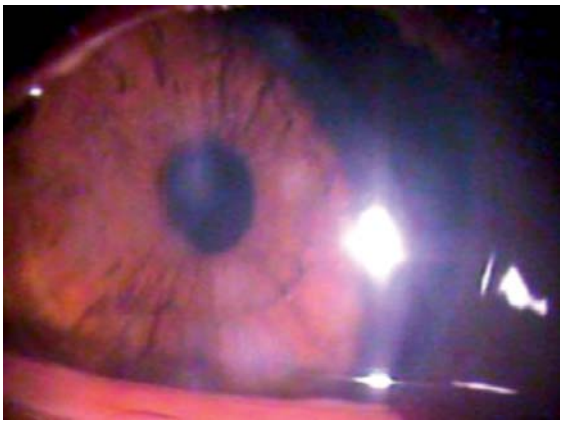

Fig. 11.9: Corneal infiltrates in epidemic keratoconjunctivitis

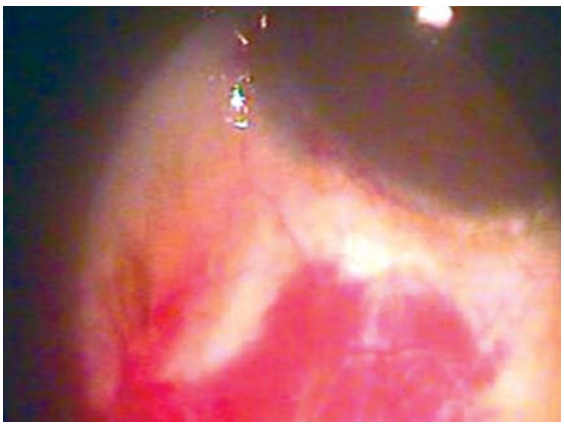

Fig. 11.10: Acute hemorrhagic conjunctivitis showing hemorrhages in the bulbar conjunctiva

lymphadenopathy are the hallmarks of the disease. Edema of the eyelids and chemosis of the conjunctiva are marked. The disease may cause transient blurring of vision.

Complications Ocular complications except punctate keratopathy are seldom seen. Neurological sequel (radiculomyelitis) is noticed in a few cases.

Treatment Acute hemorrhagic conjunctivitis has no curative treatment, it has a self-limiting course. Broad-spectrum antibiotics should be used to prevent secondary bacterial infection and cross-infection.

Chronic Conjunctivitis

Chronic conjunctivitis may occur as a legacy from an inadequately treated acute conjunctivitis or as simple chronic conjunctivitis or specific granulomatous conjunctivitis.

Simple Chronic Conjunctivitis

Simple chronic conjunctivitis is marked by congestion of the posterior conjunctival vessels and papillary hypertrophy of the palpebral conjunctiva associated with burning or grittiness in the eye.

Etiology The condition results from continuation of an acute conjunctivitis in absence of an adequate treatment. Errors of refraction, nasal or upper respiratory tract catarrh, pollution from smoke and dust, abuse of alcohol, insomnia and metabolic disorders more often than not predispose to simple chronic conjunctivitis. Occasionally, chronic dacryocystitis, rhinitis or blepharitis may be associated with it. *Staphylococcus aureus* is usually cultured from conjunctival *cul-de-sac* of these patients.

Clinical features The patient often complains of burning and heaviness of the eyes and feels difficulty in keeping the eyes open. The symptoms are usually exaggerated during evening hours. Presence of concretion, trichiasis, foreign body or dacryocystitis causes unilateral chronic conjunctivitis. White scanty discharge is deposited on the canthi due to vicarious activity of the meibomian glands.

Treatment The treatment of chronic conjunctivitis includes elimination of predisposing and causative factors. A course of topical antibiotics usually controls the infection but symptoms may persist. Astringent drops provide symptomatic relief.

Angular Conjunctivitis

Intense itching, conjunctival congestion towards the inner and outer canthi, excoriation of the skin of lid margins at the angle and scanty mucopurulent discharge characterize angular conjunctivitis.

Etiology The condition is caused by *Morax-Axenfeld* gram-negative diplobacilli (*Moraxella lacunata*), arranged end-to-end in pairs. The organism liberates a proteolytic enzyme which macerates the epithelium of the lid margin. *Staphylococci* can also cause such a condition.

Clinical features Itching, burning, discomfort, frequent blinking and slight mucopurulent discharge are common symptoms. There occurs redness of the conjunctiva towards the canthi associated with blepharitis. Shallow marginal corneal ulcers may occasionally be found.

Treatment The diplobacillary conjunctivitis responds quickly to the application of tetracycline or oxytetracycline ointment (1%) 2 to 3 times a day. Topical eye drops containing zinc (0.125-0.25%) are also effective as they inhibit the proteolytic ferment.

Follicular Conjunctivitis

The inflammatory reaction of the conjunctiva to noxious agents usually manifests in two forms—an acute generalized *papillary hyperplasia* (vascularization with epithelial hyperplasia) and

a localized aggregation of lymphocytes (*follicles*) in the subepithelial adenoid layer. It is not infrequent to observe both the reactions occurring concurrently in the diseased conjunctiva. The follicles in the conjunctiva may be found in acute conjunctivitis, chronic conjunctivitis, as a result of allergic or toxic response to the drugs such as topical atropine and pilocarpine, and in benign folliculosis of unknown etiology.

Trachoma

The word trachoma is derived from a Greek word meaning rough. Trachoma is a specific type of contagious keratoconjunctivitis of chronic evolution characterized by follicles, papillary hypertrophy of the palpebral conjunctiva, neovascularization and infiltration of the cornea (pannus) and, in late stages, conjunctival cicatrization. It is one of the oldest and most widespread diseases affecting more than one-fifth of the population of the world. It is still an important cause of visual impairment and blindness. The distribution of the disease in the world is heterogeneous. It is highly prevalent in North Africa, Middle-East and certain regions of South-East Asia. No race is immune to this disease.

It is increasingly realised that trachoma in its natural course has a low contagiousness but becomes endemic only when there exists environmental factors favoring the transmission. In trachoma endemic zones, it is almost always contacted in infancy; eye-to-eye transmission can be considered as a rule. In sporadic cases, genitals may be the source of infection. Overcrowding, abundant fly population, insanitary conditions, paucity of water and poor personal hygiene contribute to the dissemination and persistence of the infection. Trachoma seldom occurs in pure form in endemic zones where secondary bacterial or viral infections superimpose. The latter helps in transmission by increasing the conjunctival secretion and adds to the severity of the disease due to gross cicatricial sequelae.

Etiology Trachoma is caused by a large-sized atypical virus belonging to the psittacosis-lymphogranuloma-trachoma (PLT) group—*Chlamydia trachomatis*. Microimmunofluorescence test is the serologic standard for Chlamydia. As many as 14 serotypes of Chlamydia are recognized and designated by the letters A, B, Ba, C, D, Da, E, F, G, H, I, Ia, J and K. The agents isolated from the patients of trachoma and inclusion conjunctivitis are indistinguishable, hence, two are jointly known as *TRIC agent* (TR for trachoma and IC for inclusion conjunctivitis). The life cycle of the agent can be studied in the scrapings from the conjunctiva.

Life cycle of chlamydia trachomatis *Chlamydia trachomatis* forms colonies in the conjunctival epithelial cells called *Halberstaedter-Prowazek inclusion bodies* (Fig. 11.11). A few healthy epithelial cells are attacked by small *elementary bodies* which take intracellular extranuclear position. They swell to form ill-defined *initial bodies*. On staining, the initial bodies take violet stain. They rapidly divide into small, multiple elementary bodies embedded in a carbohydrate matrix to form the inclusion body, and displace the nucleus of the cell. The cell swells up and ultimately bursts to set free the elementary bodies which may attack other cells.

Pathology The TRIC agent induces papillary hyperplasia of the epithelium and lymphoid

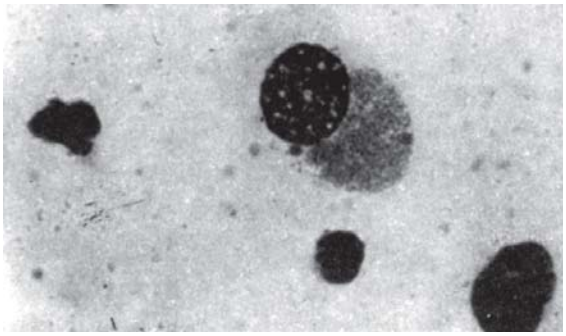

Fig. 11.11: Trachoma inclusion bodies

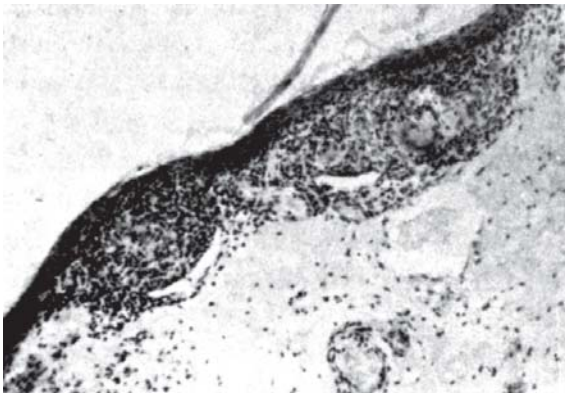

Fig. 11.12: Histopathology of trachomatous follicles

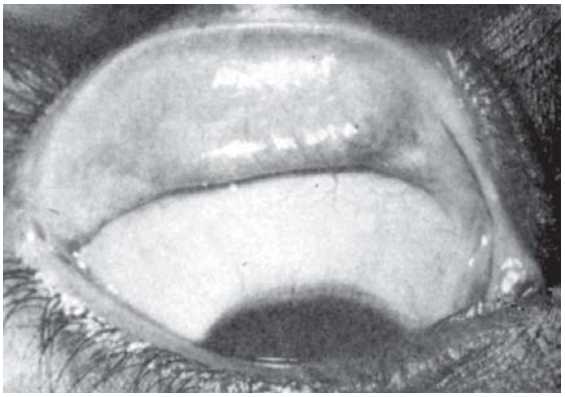

Fig. 11.13: Papillary hyperplasia of conjunctiva: Trachoma stage 1

infiltration in the adenoid layer of the conjunctiva. Localized aggregations of lymphocytes form follicles which undergo necrotizing change. The follicle (Fig. 11.12) is invaded by multinucleated macrophages (Leber's cells) which engulf the cytoplasmic and nuclear debris. At this stage, fibroblasts grow from the periphery and result in scarring. The cicatrized conjunctiva may undergo hyaline or amyloid degeneration. The necrotic and cicatricial changes in trachoma follicles distinguish them from non-trachomatous follicles as none of these changes develop in the latter.

Clinical features In most of the cases trachoma has an insidious onset after an incubation period of 5 to 15 days. In pure form it is a symptomless disease which undergoes spontaneous regression in persons with good personal hygiene.

Acute or subacute onset of trachoma is seen in adults which resembles bacterial conjunctivitis in signs and symptoms. The symptoms of trachoma include foreign body sensation, watering, itching, photophobia and redness. The infection involves both the conjunctiva and the cornea at about the same time in majority of cases.

The conjunctival signs include congestion, diffuse papillary hyperplasia (Fig. 11.13) and appearance of follicles on the mid upper tarsal conjunctiva.

Papillary hyperplasia of conjunctiva involves mainly the upper palpebral conjunctiva that appears congested, red and thickened.

Follicle is the characteristic lesion of trachoma preferentially appears on the upper palpebral conjunctiva. The follicles appear on the lower palpebral conjunctiva as well and, occasionally, on the bulbar conjunctiva. The latter is pathognomonic of the disease. The trachoma follicles are bigger in size and variable in consistency (soft in the center and firm in the periphery) as compared to the follicles of follicular conjunctivitis. They are irregularly arranged on both the upper (Fig. 11.14) and lower palpebral conjunctivae and undergo cicatrization. The follicles of

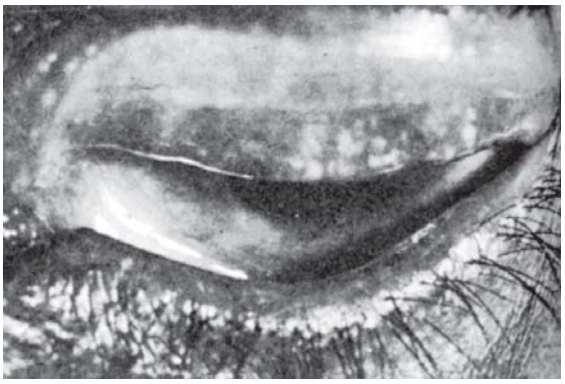

Fig. 11.14: Trachoma follicles: Trachoma stage 2

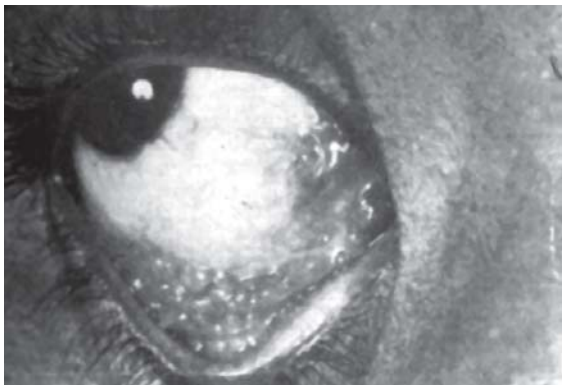

Fig. 11.15: Follicular conjunctivitis

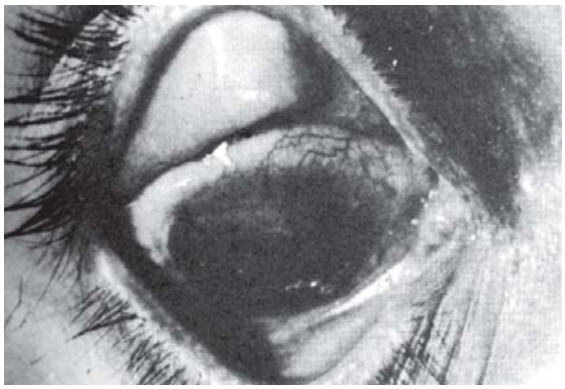

Fig. 11.16: Trachomatous pannus

follicular conjunctivitis are predominantly seen on the lower palpebral conjunctiva (Fig. 11.15), regularly arranged in rows and never undergo cicatrization.

The *trachomatous cicatrization* may be localized or diffuse. A fine linear scar appears in the sulcus subtarsalis—*Arlt's line*. Multiple star-shaped scars are seen in trachoma of moderate severity and white thick dense scarring of upper tarsal conjunctiva is commonly found in severe recurring trachoma. The latter may cause trichiasis and entropion.

The cornea is almost always involved in trachoma more or less simultaneously with the conjunctiva. Small punctate epithelial erosions over the upper half of the cornea can be demonstrated by fluorescein stain. Subepithelial infiltration may develop later. Typical follicles (Herbert's follicles) may develop on the limbus.

A superficial avascular keratitis and a thin *pannus* (lymphoid infiltration with vascularization of the upper limbus) may be evident on slit-lamp biomicroscopy in the initial stages of trachoma. However, the pannus becomes obvious with the extension of blood vessels from the vascular loops towards the center of the cornea associated with dense cellular infiltration (Fig. 11.16). In *progressive pannus*, the cellular infiltration lies beyond the terminal ends of nonanastomozing parallel vessels. But in *regressive pannus*, the vessels extend a short distance beyond the area of cellular infiltration. An extensive pannus, invading the pupillary area, causes visual impairment.

Follicles leave oval or circular pits (Herbert's peripheral pits) at the limbus (Fig. 11.17). The pits are highly pathognomonic of trachoma as none of the other ocular diseases is known to produce them.

Superficial irregular indolent ulcers may develop at the advancing edge of the pannus as a result of breakdown of pustules. They cause irritation, lacrimation and photophobia. Later, a

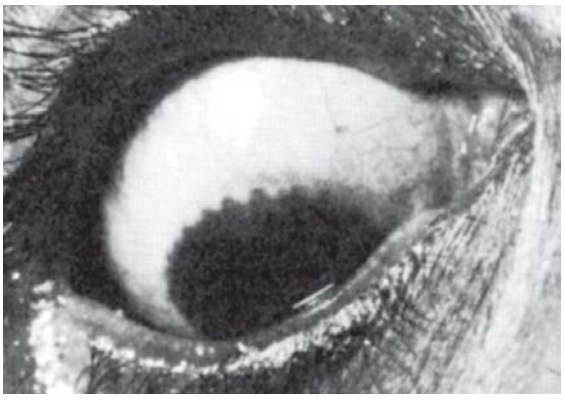

Fig. 11.17: Herbert's pit

dense corneal scar appears. In the beginning, the pannus lies between the epithelium and Bowman's membrane. Slowly, it erodes Bowman's membrane and invades the substantia propria. In such cases, resolution of pannus leaves corneal haze. However, early pannus may resolve completely without any corneal haze.

Classification The course of trachoma is arbitrarily divided into four stages by MacCallan.

Tr. I—Trachoma Stage 1 (Incipient Trachoma)

Incipient trachoma represents the earliest stage of the disease with minimal papillary hyperplasia and immature follicles on the upper palpebral conjunctiva associated with micropannus. Sometimes, clinical signs are nonconclusive and laboratory investigations like demonstration of inclusion bodies and isolation of *Chlamydia trachomatis* are required to confirm the diagnosis.

Tr. II—Trachoma Stage 2 (Manifest Trachoma)

Mature soft sagograin-like follicles in the superior tarsal conjunctiva, papillary hypertrophy, gross pannus and limbal follicles or Herbert's pits characterize this stage of trachoma.

Tr. III—Trachoma Stage 3 (Healing Trachoma)

Cicatrization or scarring develops usually around the necrotizing trachoma follicles (Fig. 11.18). Besides scarring, some or all the signs of stage 2 may be present.

Tr. IV—Trachoma Stage 4 (Healed Trachoma)

The follicles and papillary hypertrophy disappear, and the palpebral conjunctiva is completely cicatrized and smooth. The scar may be thin or dense. Pannus resolves and the presence of incomplete or complete Herbert's pits may be seen at the limbus.

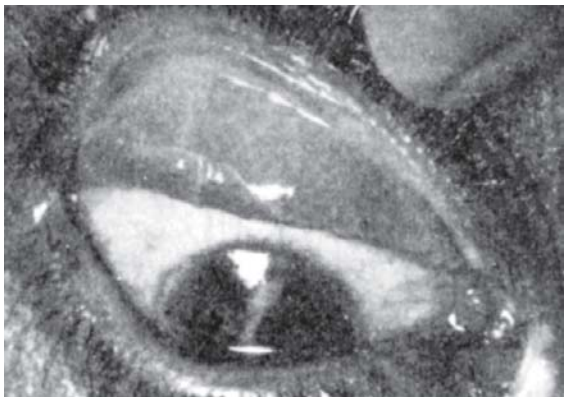

Fig. 11.18: Healing trachoma: Trachoma stage 3

The WHO has revised the classification of trachoma in 1987 mainly with the purpose of preventing the trachomatous blindness. It includes 5 stages (Table 11.3). This classification is helpful for paramedical field workers to diagnose and manage the disease.

Complications and sequelae Corneal ulceration and occasional iritis are the complications of trachoma.

In endemic zones, the disease often causes sequelae owing to cicatrization. *Trachomatous ptosis* develops following dense infiltration and cicatrization of the tarsal plate of the upper lid. The contraction of the scar tissue at the lid margin

Table 11.3: WHO classification of trachoma

Stages	*Sign*	*Definition*
TF	Trachomatous inflammation: follicular	The presence of 5 or more follicles in the upper tarsal conjunctiva
TI	Trachomatous inflammation: intense	Pronounced inflammatory thickening of the upper tarsal conjunctiva that obscures more than half the normal deep tarsal vessels
TS	Trachomatous scarring	The presence of scarring in the tarsal conjunctiva
TT	Trachomatous trichiasis	At least one eyelash rubbing on the eyeball
CO	Corneal opacity	Easily visible corneal opacity involving at least a part of pupillary margin

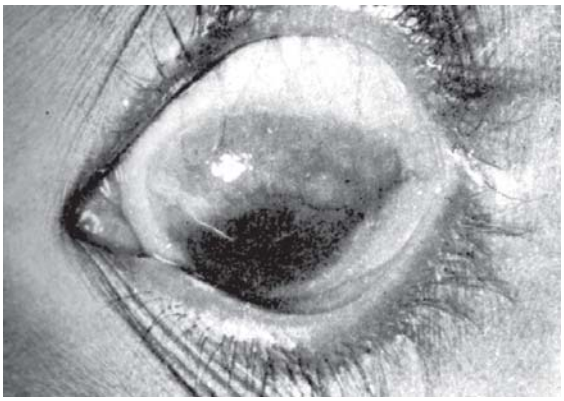

Fig. 11.19: Pannus crassus

may lead to *trichiasis* and *entropion*. Thickening of the lid margin (*tylosis*) is not uncommon. *Xerosis* and *symblepharon* may develop in the conjunctiva. *Corneal scar* and *pannus crassus* (Fig. 11.19) or *total pannus* may cause marked visual impairment and more or less total blindness. Trachomatous dacryocystitis and secondary glaucoma may occur in some patients.

Diagnosis The clinical diagnosis of trachoma requires the presence of at least two of the following signs: (i) follicles or Herbert's pits, (ii) epithelial or subepithelial keratitis, (iii) pannus, and (iv) cicatrization. The diagnosis can be confirmed by direct demonstration of the inclusion bodies in conjunctival scrapings and staining with Giemsa or iodine stain, isolation of TRIC agent and specific antibodies by microimmunofluorescence technique. DNA amplification techniques that use the polymerase chain reaction (PCR) or the ligase chain reaction (LCR) are very sensitive for diagnosing trachoma. However, these tests are time consuming and expensive.

Differential diagnosis Trachoma should be differentiated from non- trachomatous follicular conjunctivitis. Following conditions can induce follicle formation in the conjunctiva.

1. Acute follicular conjunctivitis:
   a. Inclusion conjunctivitis
   b. Adenovirus conjunctivitis:
      i. Epidemic keratoconjunctivitis
      ii. Pharyngoconjunctival fever
   c. Acute herpetic conjunctivitis
   d. Newcastle conjunctivitis
2. Chronic follicular conjunctivitis
3. Toxic follicular conjunctivitis:
   a. Miotic drugs
   b. Molluscum contagiosum
   c. Other irritants
4. Folliculosis.

Trachoma follicle can be differentiated from nontrachomatous follicle (Table 11.4). The non-trachomatous follicles preferentially develop on the lower palpebral conjunctiva and lower fornix. They are firm in consistency and never resolve by fibrosis. Out of all follicular conjunctivitis, only trachoma develops characteristic pannus.

Table 11.4: Difference between trachoma follicle and nontrachomatous follicle

	Trachoma follicle	*Nontrachomatous follicle*
Common site	Upper palpebral conjunctiva and upper fornix	Lower palpebral conjunctiva and lower fornix
Characteristics	Follicles have varying consistency often soft due to low grade necrosis	Follicles are firm in consistency
Resolution	Follicles resolve by cicatrization	Follicles resolve without cicatrization
Herbert's pits	Follicles develop at limbus and resolve by leaving characteristic Herbert's pits	Follicles do not develop at limbus and hence no pits

Inclusion Conjunctivitis

Etiology Inclusion conjunctivitis is caused by serotype D-K of *Chlamydia trachomatis*. It manifests in two forms: (i) acute papillary conjunctivitis of newborn, and (ii) acute follicular conjunctivitis of children or adults. The latter is also known as *swimming-bath* or *swimming-pool conjunctivitis*. The primary source of infection appears to be a mild urethritis in males and cervicitis in females. The transmission may occur either by fingers or through the water of the pool.

Clinical features Inclusion conjunctivitis has an acute onset. Acute follicular hypertrophy of the lower palpebral conjunctiva, mild superficial punctate keratitis or, occasional, micropannus and preauricular lymphadenopathy are the clinical features of the disease.

Treatment The disease runs a benign course. Improvement in the personal hygiene and chlorination of swimming pool check the local epidemics. Topical erythromycin 0.5% or tetracycline 1% ointment applied 4 times a day for 3 weeks provides relief. Azithromycin 1 g in a single oral dose or ofloxacin 300 mg twice a day for 1 week is effective in controlling the infection. Systemic erythromycin 500 mg 4 times a day or doxycycline 100 mg twice a day for 2 weeks may also be used.

Molluscum Contagiosum Conjunctivitis

Molluscum contagiosum is caused by a virus and it causes a low grade follicular conjunctivitis. The conjunctival lesions and corneal vascularization occur due to the release of viral proteins and other substances in the tear film. More than one molluscum nodules may be present on the lid margin (Fig. 11.20). Molluscum nodules on the skin of the eyelids are small and smooth with an umblicated core.

The treatment of toxic conjunctivitis due to molluscum contagiosum is by excision or cryo application to the eyelid nodule.

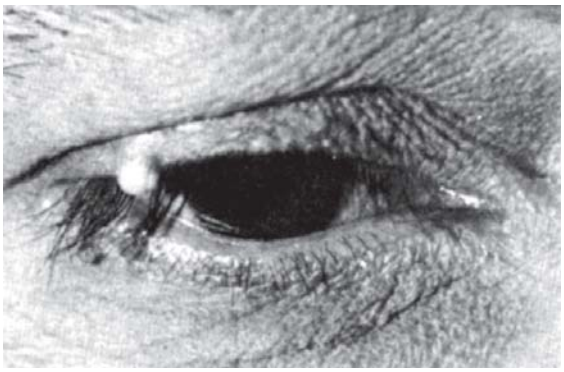

Fig. 11.20: Molluscum contagiosum

Treatment of trachoma All cases of active trachoma must be treated. Ciprofloxacin, erythromycin, tetracycline, ofloxacin and azithromycin are quite effective against TRIC agent. Chloramphenicol and penicillin are less effective. Aqueous soluble sulfonamide (20-30%) topically and long-acting sulfonamide orally may be used. However, sulfa drugs may cause allergic reaction in some patients. Instillation of ciprofloxacin 0.3% or ofloxacin 0.3% eye drop 4 times a day and application of 1% erythromycin or tetracycline ointment at bed time for 6 weeks control the infection in most cases. In addition to topical antibiotic therapy, administration of oral antibiotic (250 mg erythromycin or tetracycline 4 times a day or doxycycline 100 mg twice a day) for 3 weeks provides dramatic results. It is claimed that a single dose of azithromycin 20 mg per kg body weight for children and a single dose of 1-1.5 g for adults gives superior cure rate of trachoma. Further, azithromycin has fewer side effects than tetracycline and sulfonamides.

To combat trachomatous blindness, the WHO has developed the SAFE strategy. It is an acronym for:

S: Surgery for trichiasis
A: Antibiotic treatment of active infection
F: Facial cleanliness
E: Environmental improvement

To eliminate trachoma and its blindness each component of the SAFE strategy must be implemented.

A six-week treatment eliminates the infection from the conjunctival sac though the follicle may

not resolve. A follow-up examination is necessary to assess the complete cure of the disease. Persistent trachoma follicles were dealt with, in the past, by mechanical expression by roller forceps or by painting with copper sulphate or silver nitrate solution. Such drastic procedures resulted in heavy cicatrization, therefore, discarded. Presently, a combination of local and systemic antibiotic therapy is preferred no matter one has to continue the drug for a longer time. The management of trichiasis and entropion requires surgical intervention.

Trachoma control Trachoma is a specific communicable keratoconjunctivitis which is a public health problem in the developing countries. The disease is closely associated with personal hygiene and environmental sanitation. Trachoma often spreads by the transfer of infected conjunctival secretions through fingers, common towel and flies. Therefore, mothers are instructed not to apply eye cosmetics *(Kajal)* to all children of the family with the same finger. Free mixing of acute cases of trachoma in school or other public places should be checked strictly. Breeding of flies be minimized by adopting proper sanitary measures.

Health education on trachoma should be given to the general public. Adoptation of adequate health measures has minimized the intensity and severity of the disease even in the trachoma endemic zones. A community having more than 50% prevalence of trachoma is covered by a blanket antibiotic therapy (WHO intermittent schedule of treatment). The antibiotic ointment is applied to the entire population twice daily for 3 to 5 days in a month for 3 to 6 months. As trachoma infection does not give any lasting immunity, immunization of the population is futile. Although the trachoma control programs are being in operation in many countries, the ultimate solution of the problem lies in the overall improvement in the standard of living of trachoma affected population.

Granulomatous Conjunctivitis

Granulomatous infections such as tuberculosis, syphilis and leprosy produce specific reactions in the conjunctiva.

Tuberculosis of the Conjunctiva

Etiology Tuberculosis of the conjunctiva is uncommon, and occurs in young people. It may or may not be associated with systemic tuberculosis. The infection is usually exogenous in origin.

Clinical features The conjunctiva may rarely get infected by *Mycobacterium tuberculosis*. The infection is invariably exogenous in origin. The preauricular lymph glands are often involved and tend to suppurate.

Types of lesions The tubercular lesions of the conjunctiva may manifest in following forms:

1. Small multiple miliary ulcers on the palpebral conjunctiva
2. Granular or follicular type of conjunctivitis
3. Gelatinous cock's comb-like excrescences in the fornices
4. Polypoid pedunculated outgrowth
5. Tubercular nodule at the limbus.

Pathology Histopathology of the lesion presents a typical tubercle formation with Langerhan's giant cells. The conjunctival scrapings may show acid-fast tubercular bacilli.

Treatment The primary affection of the conjunctiva requires excision and cauterization. However, a complete course of systemic antitubercular drugs should be administered.

Syphilitic Conjunctivitis

Syphilitic lesions of the conjunctiva are uncommon. Conjunctiva may be affected in all the three stages of the disease.

A *primary chancre* may rarely develop in the conjunctiva. It may resemble a chalazion if present on the palpebral conjunctiva.

A *catarrhal conjunctivitis* may occur in the secondary stage of syphilis.

A *gumma* or *gummatous ulceration* of the conjunctiva associated with enlarged preauricular lymph glands may be found in the tertiary syphilis.

Besides syphilis and tuberculosis, conjunctival ulceration may occur due to trachoma and foreign body.

Diagnosis The demonstration of spirochetes in the scraping from the lesion and positive fluorescent treponemal antibody absorption (FTA-ABS) test confirm the diagnosis.

Treatment A full course of systemic antisyphilitic drugs and topical tetracycline should be administered.

Leprotic Conjunctivitis

Ocular involvement in leprosy is not infrequent. Nonspecific conjunctivitis may develop. There may be nodules on the lids, limbus or cornea. Exposure keratitis consequent to Bell's palsy occurs in late cases of leprosy.

Parinaud Oculoglandular Syndrome

Parinaud oculoglandular syndrome (POS) is characterized by follicular conjunctivitis with regional lymphadenopathy and symptoms of fever, headache and anorexia. Rarely, the disease may cause optic neuritis, encephalitis and hepatitis.

Etiology The disease is mainly caused by *Bartonella henselae* (cat-scratch disease); other causes include tularemia, tuberculosis, syphilis, sarcoidosis and fungal infections.

Treatment Currently no definitive treatment is available. Azithromycin, ciprofloxacin, erythromycin or doxycycline may be tried systemically along with NSAIDs.

Fungal Conjunctivitis

Candida albicans, *Nocardia*, *Aspergillus* and *Sporothrix* can cause chronic conjunctivitis. *Candida* in debilitated persons may produce a pseudomembranous or ulcerative conjunctivitis. *Leptothrix* and other fungi may cause follicular conjunctivitis associated with preauricular lymphadenopathy. Topical fluconazole or miconazole 1% and natamycin are used in the treatment of fungal conjunctivitis.

Rhinosporidiosis of the Conjunctiva

Rhinosporidiosis of the conjunctiva is not a rare fungal affection of the conjunctiva. It is caused by *Rhinosporidium seeberi*. The characteristic conjunctival lesions are pedunculated or sessile fleshy growths with irregular surface dotted with white spots (Fig. 11.21). The effective treatment is complete surgical removal of the growth.

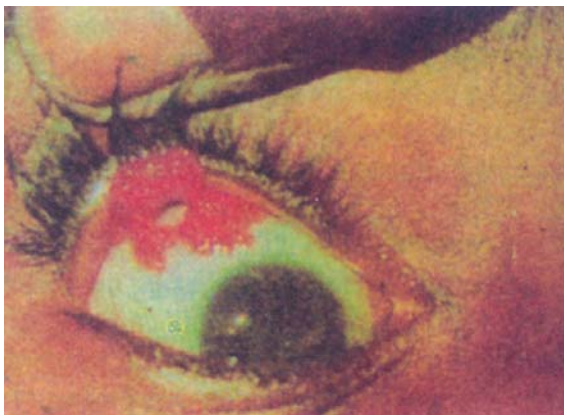

Fig. 11.21: Rhinosporidiosis of conjunctiva
(*Courtesy:* Dr TP Itteyrah, Little Flower Hospital, Angamally)

Ophthalmia Nodosa

Ophthalmia nodosa is a foreign body nodular conjunctivitis caused by the retained hair of caterpillars. The condition is common in summer months and the lesion consists of yellowish-gray translucent raised nodule on the bulbar conjunctiva.

The nodule is formed as a result of lymphocytic and giant cells infiltration around the hair. Excision of the nodule gives relief.

Oculocutaneous Syndromes

Inflammation of conjunctiva, inflammatory involvement of mucous membranes of mouth, nose, urethra and vulva, eruptive lesions of the skin, and varying degree of constitutional symptoms are found in a number of clinical entities (Stevens-Johnson syndrome, Reiter's syndrome and Behçet's syndrome) described under *erythema multiforme* or oculocutaneous syndrome. Pemphigus or pemphigoid reaction in the conjunctiva is rare and also included in the oculocutaneous syndrome.

Numerous vesicles appear on the conjunctival surface, they rupture and undergo progressive cicatrization causing *essential shrinkage of the conjunctiva* often associated with corneal complications and xerophthalmia.

Treatment of oculocutaneous syndrome is unsatisfactory. However, artificial tears and grafting of amniotic membrane and stem cells transplantation by means of conjunctival autograting may be helpful.

Allergic Conjunctivitis

Allergic or hypersensitivity reactions of the conjunctiva are not uncommon. They may be immediate (humoral) as seen in hay fever, acute or subacute conjunctivitis and vernal conjunctivitis, or delayed (cellular) as found in phlyctenular conjunctivitis.

Acute or Subacute Allergic Conjunctivitis

Acute or subacute catarrhal inflammation of conjunctiva is often associated with allergic rhinitis.

Etiology The condition is caused by exogenous allergens such as pollen, grass, animal dander, etc. Occasionally, cosmetics, chemicals and drugs applied topically can induce a violent follicular or nonfollicular reaction in the conjunctiva. The conjunctivitis is often seen in the Western countries as a part of typical hay fever, hence, known as *hay fever conjunctivitis*.

Clinical features Itching, watering and redness of the eye are common complaints of the patient. Mild to moderate injection of the conjunctiva and severe chemosis are found. Scrapings from the conjunctiva show some eosinophils. Remissions are common.

Treatment The disease can be prevented by the elimination of allergens from the surroundings or the patient may be moved to a pollen-free area. Desensitization against specific allergen may be helpful but is a cumbersome process. Symptomatic relief is quickly obtained by cold compresses and instillation of corticosteroid drops. Astringent lotions and antihistaminic drops bring temporary relief. Cromolyn sodium 2-4% drops 4 times a day and olopatadine hydrochloride 0.1% drops 2 times a day are effective in controlling the seasonal exacerbations. Relief from itching may be obtained by giving systemic antihistaminics.

Vernal Keratoconjunctivitis

Vernal keratoconjunctivitis (VKC), a recurrent bilateral seasonal conjunctivitis, is characterized by intense itching, photophobia, white ropy discharge and appearance of well-defined polygonal raised areas of papillary hypertrophy on the palpebral conjunctiva and a wall of gelatinous thickening at the limbus.

Etiology Vernal keratoconjunctivitis is caused by an immediate hypersensitivity reaction to some exogenous allergens. The immunopathogenesis involves both type I and type IV hypersensitivity reactions. VKC is found mostly in families with a history of atopy and asthma. There is an increased IgE and eosinophils in the blood. The disease has the onset in summer months, hence, it is also known as *spring catarrh*, which is a misnomer may be seen round the year in tropical climate. The disease is less common in temperate zones and almost non-existent in cold climate.

Clinical features Vernal conjunctivitis frequently affects children between 4 and 15 years, often boys more than girls. The disease shows exacerbations and remissions with change of weather. However, it is a self-limiting disease and the frequency of attacks and severity of the symptoms eventually subside as the patient ages.

The disease is usually seen in two clinical forms, the *palpebral* and the *limbal*, both may co-exist in a patient.

The *palpebral form* is relatively more common, the upper palpebral conjunctiva is hypertrophic and shows the presence of small to giant papillae (Fig. 11.22). Each papilla is polygonal with a flat top and contains tufts of capillaries and dense fibrous tissue. The polygonal raised areas of palpebral conjunctival hypertrophy are seen mimicking cobblestones. The hyaline degeneration imparts bluish-white or milky color to the papilla. The papillae may also appear in the lower palpebral conjunctiva. A stringy conjunctival discharge or a fibrinous pseudomembrane (Maxwell-Lyons sign) may sometimes be found.

The *limbal* or *bulbar form* is less characteristic and frequently seen in black races. The striking lesion is at the limbus where a wall of gelatinous thickening appears (Fig. 11.23). It may be associated with micropannus (Fig. 11.24). As the disease progresses, the thickening becomes irregular and a few discrete, white, superficial

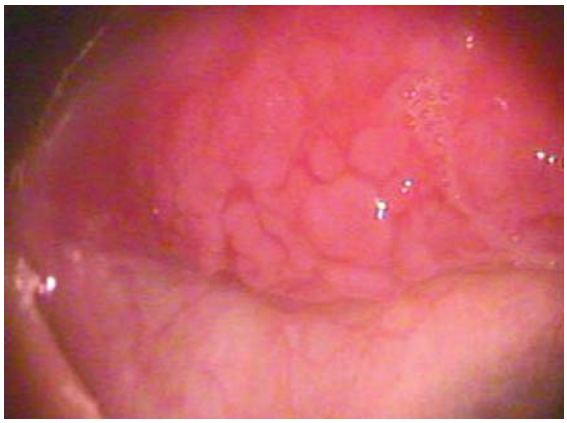

Fig. 11.22: Palpebral form of vernal conjunctivitis: Moderate papillae

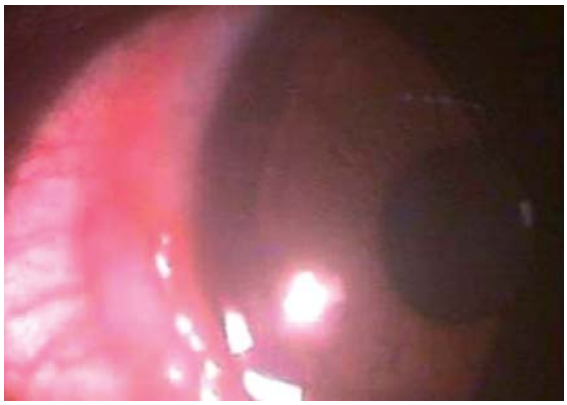

Fig. 11.23: Bulbar form of vernal conjunctivitis

Fig. 11.24: Vernal conjunctivitis: Micropannus and corneal infiltrates

dots or nodules, *Horner-Trantas' spots,* appear at the limbus that are mainly composed of degenerated eosinophils.

The corneal lesions of vernal conjunctivitis (*vernal keratopathy*) include superficial punctate keratitis, epithelial erosions, noninfectious oval ulcer (shield ulcer), subepithelial scarring and pseudogerontoxon with a classical cupid-bow outline. An association between VKC and keratoconus has been reported.

Pathology Smears of conjunctiva show the presence of eosinophilic granules in great numbers. Histopathology of vernal conjunctivitis reveals: (i) excessive epithelial hyperplasia, (ii) extensive infiltration by eosinophils, plasma cells, lymphocytes and monocytes in the adenoid layer, and (iii) spectacular increase in the fibrous layer which later on undergoes hyaline degeneration.

Treatment

Mild to moderate VKC: Topical cromolyn sodium and ketorolac tromethamine offer relief in patients with year-round disease. Diclofenac sodium or ketorolac tromethamine 0.5% drops are considered safe and may be used on a long-term basis. In mild to moderate symptom-free cases only cromolyn sodium is needed. Lodoxamide tromethamine (0.1% solution 4 times daily) acts faster than cromolyn sodium and relieves symptoms by reducing mast cell degranulation and inhibiting eosinophil chemotaxis. Photophobia in VKC can be prevented by wearing dark glasses.

Severe VKC: Severe cases or patients with seasonal exacerbation need topical corticosteroids. The instillation of corticosteroids should be tapered gradually. To avoid corticosteroid-related complications, intermittent (pulse) therapy is indicated. Soluble corticosteroid drops are used 2-4 hourly for 5-7 days and then rapidly tapered. An alternative to topical steroid therapy is an injection of triamcinolone acetonide (40 mg per ml) injected in the supratarsal conjunctiva.

Refractory VKC: Refractory cases of VKC usually do not respond to routine therapy. An immunosuppressive agent, cyclosporin A, that inhibits chemotaxis, can be used as 1-2% drops in these cases.

Giant papillae are treated either by application of β-radiation (600-1500 rad) or by cryo application. Persistent giant nodules need excision.

Giant Papillary Conjunctivitis

A giant papillary reaction in the conjunctiva occurs in contact lens (hydrophilic lenses) wearers, patients with ocular prosthesis and patients in whom corneal sutures, particularly of keratoplasty, are not removed. The papillae are polygonal and resemble cobblestones as in vernal conjunctivitis. They are composed of eosinophils, basophils and mast cells.

Local corticosteroid drops or cromolyn sodium drops may relieve the symptoms of foreign body sensation, itching and photophobia transiently.

Cleaning of the deposits on the contact lens, discontinuation of lens or prosthesis wear and removal of corneal sutures are effective measures to manage the papillae.

Phlyctenular Conjunctivitis

Phlyctenular conjunctivitis is an endogenous allergic conjunctivitis marked by photophobia, mucopurulent discharge and presence of a single or multiple gray-white raised nodules at the limbus surrounded by an area of conjunctival congestion.

Etiology Phlyctenular conjunctivitis is a delayed hypersensitivity (type IV, cell-mediated) response to endogenous microbial proteins which in most of the cases are tubercular or staphylococcal. Phlyctenulosis may occur secondary to staphylococcal blepharitis. The disease is common in malnourished and debilitated children between 5 and 12 years of age. These children suffer from enlarged tonsils and cervical lymphadenopathy.

Clinical features Phlyctenular conjunctivitis is usually unilateral, but the other eye may get involved in a few months. The disease in a pure form does not give many symptoms except mild discomfort and irritation with reflex lacrimation. However, as the disease is usually complicated by mucopurulent conjunctivitis, photophobia and mucopurulent discharge become prominent symptoms.

The characteristic lesion of the conjunctivitis is a *phlycten* or *phlyctens* (blebs). Single or multiple, small, round white or gray nodules raised above the surface are found at or near the limbus (Fig. 11.25A). The presence of phlycten on the palpebral conjunctiva is a rarity. The size of the phlycten may vary from 0.5 to 4 mm. The bigger phlycten appears as a pustule (Fig. 11.25B) and overlying epithelium undergoes ulceration. Both vascular and cellular reactions occur around the phlycten.

The cornea is infiltrated or may be invaded by a corneal phlycten. Corneal phlycten often causes pain and photophobia. A pannus is seen around a raised gray phlycten. The phlycten ulcerates and forms a triangular fascicular ulcer with prominent vascularization. Multiple phlyctens may surround the cornea and their subsequent necrosis leads to the formation of a *ring ulcer*. The phlycten resolves by cicatrization, in cornea the scar undergoes nodular dystrophy.

Pathology The histopathology of a phlycten shows a characteristic subepithelial mononuclear infiltration in a triangular area, the apex of the triangle being towards the deeper layers of the cornea. When secondary infection supervenes, additional polymorphonuclear cells appear and the overlying epithelium undergoes necrosis.

Differential diagnosis Besides phlycten, a nodule at the limbus may be seen in episcleritis, inflamed pinguecula, filtering bleb following glaucoma surgery, suture cyst, dermoid and foreign body granuloma. The distinguishing features of phlyctenular conjunctivitis, inflamed pinguecula and episcleritis are listed in Table 11.5.

Treatment The treatment of phlyctenular conjunctivitis is aimed to improve the general health of the child and management of local condition. Infected tonsils and adenoids should be properly treated and attempts should be made to desensitize the patient against tubercular or Staphylococcal allergens. A calorie-rich diet supplemented with vitamins A, C and D and calcium should be given. Concurrent infections need systemic antibiotic therapy.

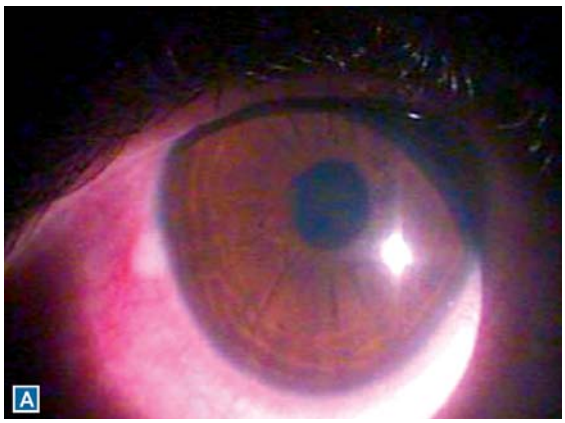

Fig. 11.25A: Phlyctenular conjunctivitis

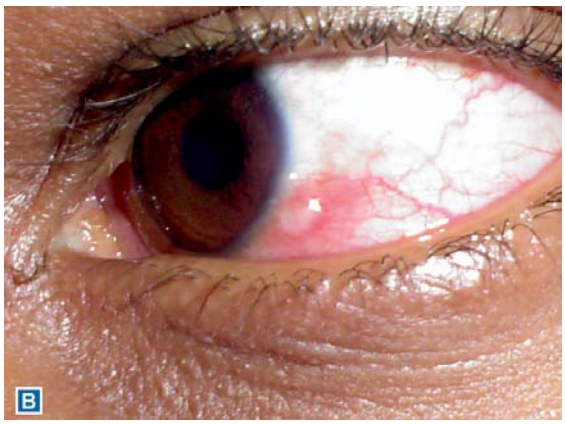

Fig. 11.25B: A big phlycten appearing as pustule (*Courtesy:* Prof. Manoj Shukla and Dr Prashant Shukla, AMUIO, Aligarh)

Table 11.5: Distinguishing features of phlyctenular conjunctivitis, inflamed pinguecula and episcleritis

Features	*Phlyctenular conjunctivitis*	*Inflamed pinguecula*	*Episcleritis*
1. Age	Below 15 years (Children)	Above 50 years (Elderly)	16-40 years (Young)
2. Sex	Both sexes	Male	Female
3. Site	Usually at limbus	Away from limbus and usually nasally	Away from limbus and usually temporally
4. Shape	Small raised round nodule	Flat and triangular	Relatively bigger flat round nodule
5. Color	White	Dull-pink, fleshy	Dull purple
6. Discharge	Mucopurulent	No discharge	Watery
7. Ulceration	Common	Does not occur	Does not occur
8. Tenderness	Seldom	May be present	Usually present
9. Regional lymph glands	Enlarged	Not enlarged	Enlarged
10. Complications	Keratitis and nodular degeneration	May lead to pterygium formation	Rarely scleritis

Hot compresses and irrigation with saline reduce mucopurulent discharge. Instillation of antibiotic and corticosteroid eye drop several times in a day has a dramatic effect on the secondary infection as well as on the phlycten. The latter disappears within a week. When cornea is involved cycloplegic should be applied. Recurrences are frequent if general condition is not dealt with. Tinted glasses protect against glare and photophobia.

Toxic Conjunctivitis

Etiology Toxic conjunctivits may occur following the use of ophthalmic medications. Benzalkonium chloride and thiomersal are often used as preservatives in ophthalmic preparations which may cause toxic reaction. Atropine, miotics, antiviral agents, aminoglycosides, epinephrine and apraclonidine have been associated with follicular conjunctivitis. Prostaglandin analogue and brimonidine may also cause toxic conjunctivitis.

Clinical features The characteristic toxic reaction in the conjunctiva occurs either in the form of a papillary hypertrophy or a chronic follicular conjunctivitis. Follicles are most predominantly seen on the inferior tarsal conjunctiva and fornix. Occasionally, a progressive conjunctival scarring can lead to contraction of fornices (pseudopemphigoid reaction). The corneal toxicity manifests as punctate epithelial erosion of inferior cornea or a whorl-pattern keratopathy. Rarely, stromal opacities and neovascularization may occur.

Treatment The drug should be immediately discontinued. Use of preservative-free drugs and topical lubricant drops may provide relief.

DEGENERATIVE CHANGES IN THE CONJUNCTIVA

The common degenerative conditions of the conjunctiva include concretions, pinguecula and pterygium.

Concretions

Concretions or lithiasis are small, hard, elevated yellow deposits in the palpebral conjunctiva. They never undergo calcareous degeneration, therefore, the term is a misnomer. Concretions are caused by

accumulation of inspissated mucus and degenerated epithelial cells in the loops of Henle. They are commonly found in elderly persons suffering from trachoma or chronic conjunctivitis. Foreign body sensation is the main symptom and occasionally a corneal abrasion may develop. Removal of concretions with a sharp needle after topical anesthesia eliminates the symptoms.

Pinguecula

Pinguecula is a degeneration of the bulbar conjunctiva characterized by the presence of a yellowish triangular spot near the limbus, the apex of the triangle being towards the canthus. It appears on the nasal side first and then the temporal side is affected. The condition is found in elderly people, especially in those living in dusty and windy climate. The name pinguecula is derived from *pinguis* meaning fat. However, it is really a hyaline infiltration and elastotic degeneration of the submucosa of the conjunctiva with little or no vascularization. It is usually stationary and does not need treatment. If pinguecula causes cosmetic disfigurement it has to be surgically removed.

Pterygium

The term pterygium is derived from the Latin word meaning wing. It is characterized by a triangular encroachment of the conjunctiva onto the cornea usually on the nasal side.

Etiology Etiology of pterygium is disputed. A number of theories such as primary degeneration of the conjunctiva and the cornea (Fuchs), inflammatory response of the conjunctiva (Kamel) and irritative reaction to ultraviolet (UV) light have been propagated. Currently, pterygium is believed to be a growth disorder characterized by conjunctivalization of the cornea due to localized UV rays induced damage to the limbal stem cells. Pterygium is common in sunny, dusty, sandy or wind blown areas of Australia, Middle-East, South-Africa and Texas, and represents a response to chronic dryness and UV exposure.

Pathologic changes in the conjunctiva are basically the same as those in the pinguecula, but proliferative inflammatory reaction is quite prominent. The vascularized granulation tissue invades Bowman's membrane and superficial layers of the corneal stroma.

Clinical features Pterygium seldom gives any symptom but its progression may cause astigmatism and its extension in the pupillary area of the cornea may cause serious visual impairment.

Classically, a pterygium has four parts: (i) a blunt apex, *head*, (ii) a few infiltrates in front of the apex, *cap*, (iii) a limbal part, *neck,* and (iv) a bulbar portion extending between the limbus and the canthus, *body*.

A *progressive pterygium* is thick and vascular (Fig. 11.26) and encroaches onto the cornea with prominent infiltrates. *Stocker's line* represents deposition of iron in the corneal epithelium anterior to the head of the pterygium. When pterygium stops growing, infiltration and vascularization disappear, and it becomes pale and thin (Fig. 11.27).

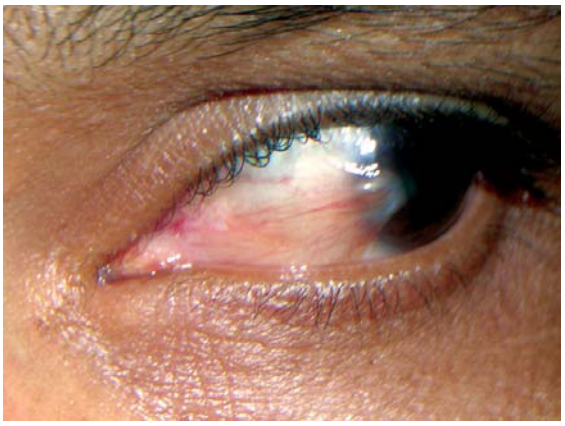

Fig. 11.26: Progressive pterygium

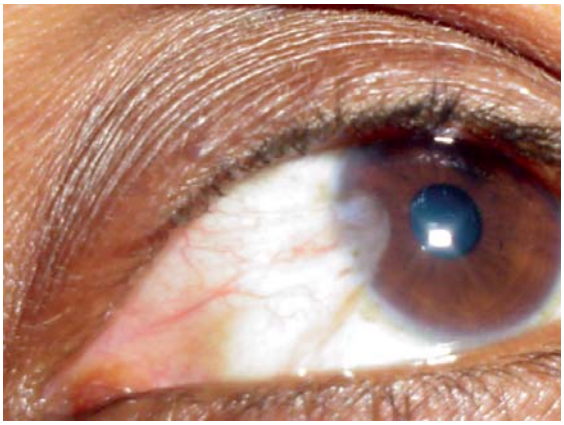

Fig. 11.27: Stationary pterygium

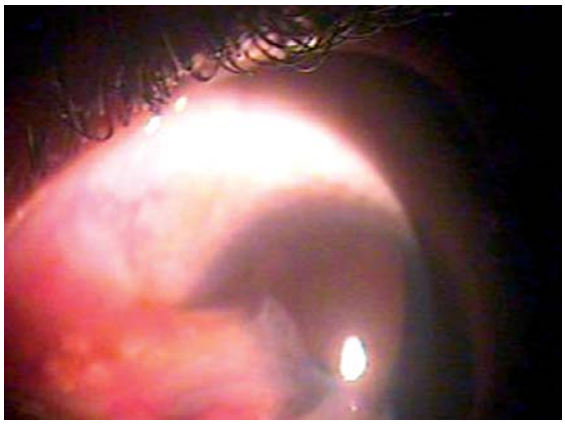

Fig. 11.28: Pseudopterygium

Treatment Pterygium requires surgical removal, especially if it threatens to encroach onto the pupillary area. Excision of pterygium is generally recommended. However, recurrence of the pterygium after surgery is not rare. An autoconjunctival graft or amniotic membrane transplantation (See video) often prevents the recurrence of pterygium. β-radiation and topical use of mitomycin-C (MMC) are also helpful in preventing the recurrence. Topical MMC may cause late aseptic scleral necrosis and sclerokeratitis in some patients.

Pseudopterygium

A pterygium-like condition (Fig. 11.28) may develop due to adhesion of the chemotic bulbar conjunctiva to a marginal corneal ulcer following acute conjunctivitis or chemical burn of the eye. It can be differentiated from a true pterygium by the passage of a probe between it and the bulbar conjunctiva.

An early age of onset, obliquity of the axis and stationary course are other differentiating features (Table 11.6). The pseudopterygium should be excised.

Table 11.6: Differentiating features between pterygium and pseudopterygium

Features	*Pterygium*	*Pseudopterygium*
1. Age	Usually seen in adults	Usually seen in children
2. Etiology	Unknown	Postinflammatory
3. Site	Palpebral aperture	Palpebral aperture cum fornix
4. Configuration	Horizontal	Oblique
5. Status	Progressive or stationary	Almost always stationary
6. Neck	Adherent to limbus	Free
7. Probe	Cannot be passed underneath	Can be passed

CYSTS AND TUMORS OF THE CONJUNCTIVA

Cysts of the Conjunctiva

The conjunctiva is a common site for development of cysts. Conjunctival cysts may occur due to dilatation of lymph spaces, *lymphangiectasis*, of the bulbar conjunctiva. Sometimes, a solitary multilocular cyst, *lymphangioma*, (Fig. 11.29) may be found. Occasionally, obstruction of the duct of

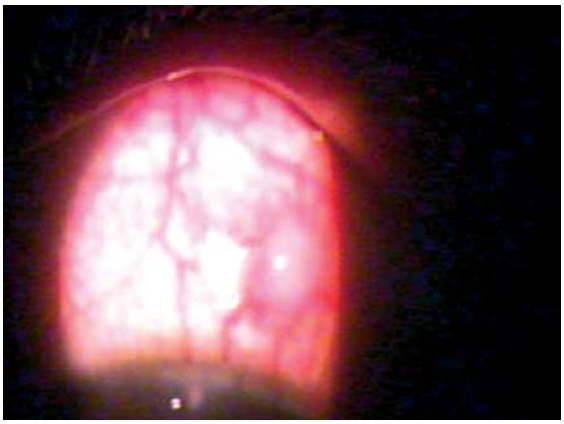

Fig. 11.29: Lymphangioma of conjunctiva

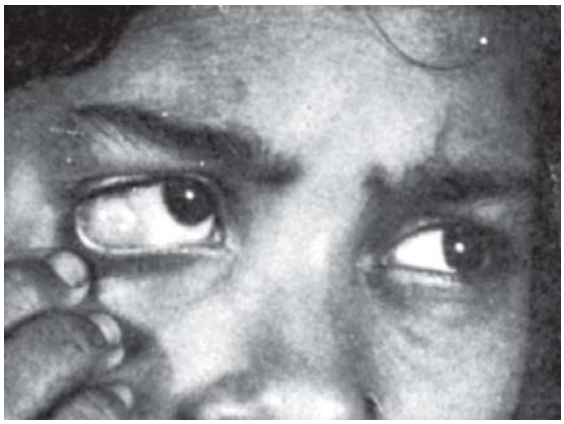

Fig. 11.31: Cysticercus cyst of conjunctiva

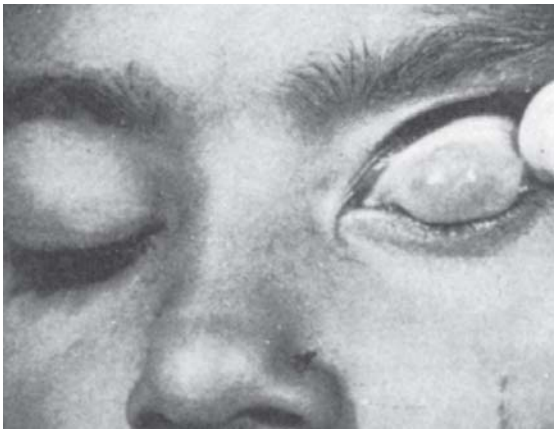

Fig. 11.30: Retention cyst of upper palpebral conjunctiva

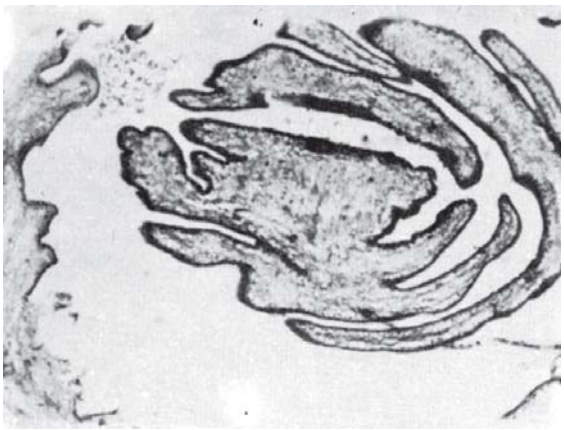

Fig. 11.32: Histopathology of cysticercus

Krause's accessory lacrimal gland results in a large *retention cyst* (Fig. 11.30). *Implantation cysts* may develop following strabismus surgery or injury. Subconjunctival *cysticercus* (Figs 11.31 and 11.32) and *hydatid cysts* are not rare in the developing countries. They require careful surgical removal.

Congenital Epibulbar Dermoid

Congenital epibulbar dermoid of the conjunctiva is a white or yellow oval mass at the limbus covering partly the conjunctiva and partly the cornea. Conjunctival dermoid (Figs 11.33 and 11.34) grows slowly and consists of epidermoid epithelium and fibrous tissue containing hair follicles and sebaceous glands.

Dermolipoma

Dermolipoma is a yellowish-white, fibro-fatty congenital tumor commonly found at the outer canthus.

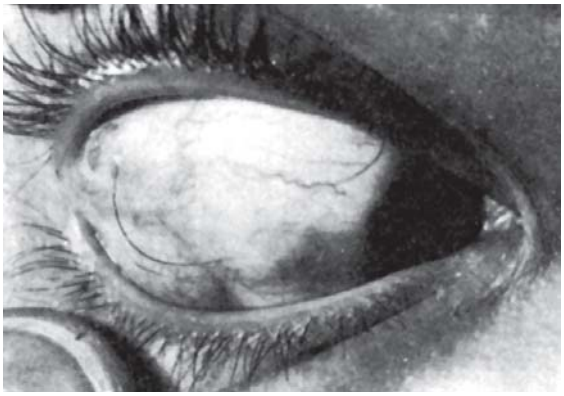

Fig. 11.33: Dermoid of conjunctiva

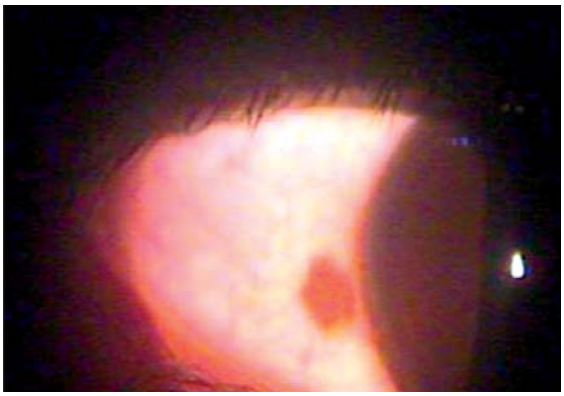

Fig. 11.35: Nevus of conjunctiva

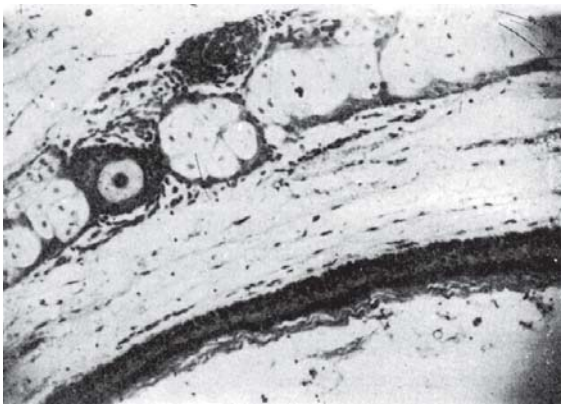

Fig. 11.34: Histopathology of dermoid of conjunctiva

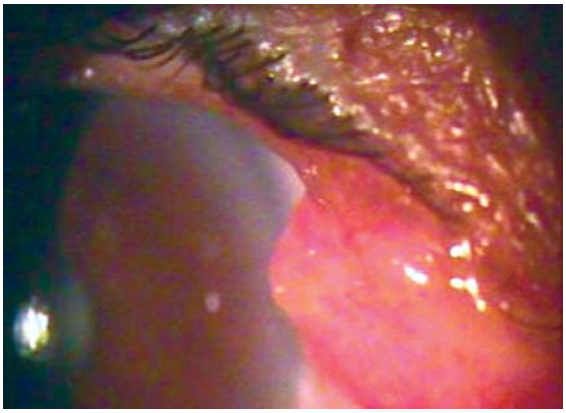

Fig. 11.36: Papilloma of conjunctiva

Tumors of the Conjunctiva

Both benign and malignant tumors may involve the conjunctiva.

Benign Tumors

Nevus

Nevus or congenital mole is frequent on the bulbar conjunctiva (Fig. 11.35). It is congenital and tends to grow at puberty. It appears as brownish or black flat lesion. Histologically, it is composed of nests of typical pigmented nevus cells. The nevus does not require excision lest malignant changes develop.

Papilloma

Papilloma is a benign polypoid tumor of the conjunctiva occurring in the fornix or at the canthus (Fig. 11.36). It may resemble the cock's comb type of tuberculosis of the conjunctiva. As it has a tendency to become malignant, it should be removed.

Fibroma

Fibroma is a rare firm or hard polypoid growth often seen in the lower fornix needing surgical removal.

Angiomas

Conjunctival angiomas are congenital and may be hemangioma or lymphangioma. Hemangiomas manifest as capillary nevi or encapsulated hemangioma. The latter is more common.

Granuloma

Granuloma of the conjunctiva may develop either on the palpebral or on the bulbar conjunctiva (Fig. 11.37) The granuloma may develop following strabismus surgery, retained foreign body and extrusion of chalazion through the conjunctiva. It may appear as a cauliflower-like (Fig. 11.38) or fungating mass of granulation tissue. Granuloma often needs surgical removal.

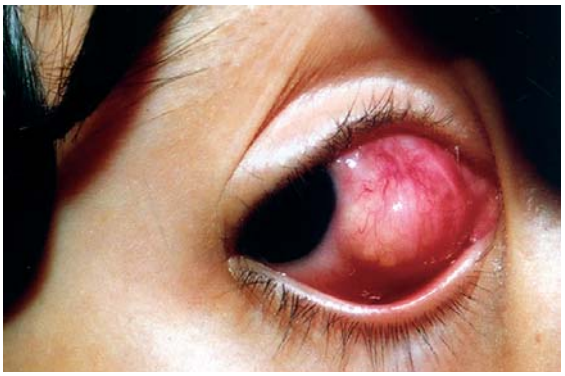

Fig. 11.37: Granuloma of bulbar conjunctiva

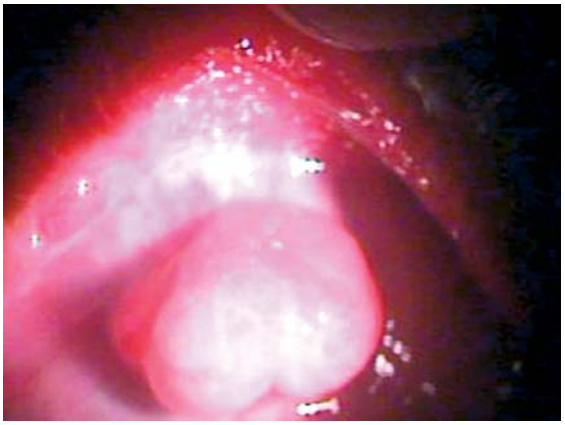

Fig. 11.38: Granuloma of palpebral conjunctiva

Malignant Tumors

Squamous Cell Carcinoma

Squamous cell carcinoma or epithelioma (Fig. 11.39) is a fleshy vascular gelatinous mass with feeder vessels usually seen at the limbus or at the lid margins. Treatment includes surgical excision with adjunctive cryotherapy or topical MMC. Intraocular spread of tumor warrants enucleation of the eyeball.

Intraepithelial Epithelioma

Intraepithelial epithelioma or Bowen's carcinoma is a rare epibulbar tumor with low malignant potential. Epithelioma can involve an extensive conjunctival area and may rarely cause perforation of the globe. Treatment consists of free excision of conjunctiva with adjunctive cryotherapy or topical MMC or 5-fluorouracil to avoid recurrence.

Basal Cell Carcinoma

Basal cell carcinoma is a common tumor which usually involves the plica semilunaris and medial part of the lower lid. Surgical excision and radiotherapy are common modes of treatment.

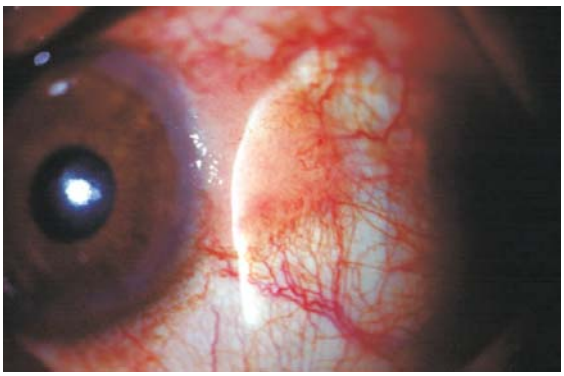

Fig.11.39: Squamous cell carcinoma of conjunctiva (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

Precancerous Melanosis

Precancerous melanosis of the conjunctiva is a diffuse pigmentation of the conjunctiva and periorbital skin (Fig. 11.40) in elderly persons. It is prone for malignancy.

Malignant Melanoma

Malignant melanoma may arise from a pre-existing nevus or *de novo* in the normal conjunctiva. It is mostly seen at the limbus and may involve other parts of the conjunctiva as well (Fig. 11.41). Metastases occur elsewhere in the body, commonly in liver. Excision of the globe or exenteration of the orbit may be required.

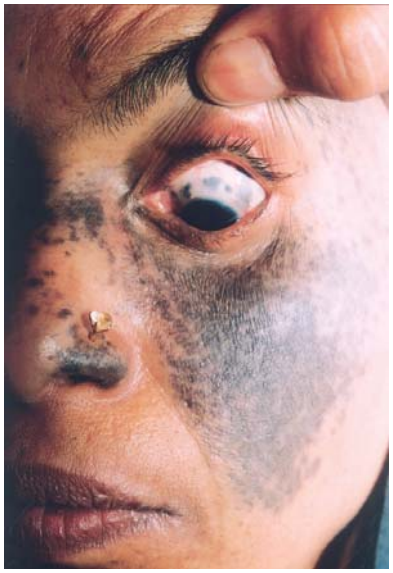

Fig. 11.40: Oculodermal melanosis

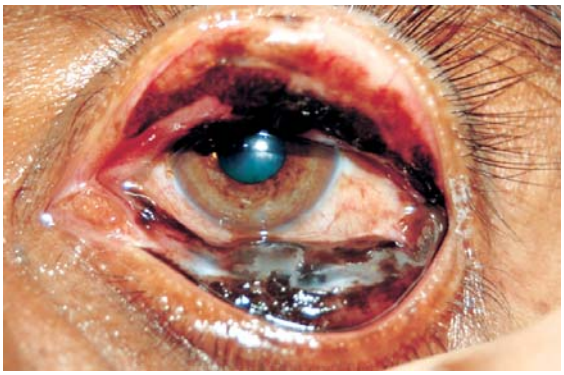

Fig. 11.41: Melanoma of conjunctiva
(*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

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CHAPTER

12 Diseases of the Cornea

ANATOMY

Cornea is a transparent avascular tissue with smooth surface (Fig. 12.1). It appears elliptical from front, its horizontal diameter being 11.5 mm and vertical about 11 mm. From the back it is, however, circular with a diameter of 11.5 mm. The cornea is thicker at the periphery (0.67 mm) than at the center (0.52 mm). The radii of curvature of the anterior and posterior surfaces of the central part of the cornea are 7.8 mm and 7 mm respectively. The cornea acts as a protective membrane as well as a strong refracting surface. It has a refractive power of about + 40 D.

The transparency of the cornea is due to its peculiar lamellar arrangement of collagen fibers, selective permeability of the epithelium and the endothelium, avascularity and deturgescence. The corneal deturgescence is maintained by an active sodium-potassium pump situated in the endothelium.

Histologically, cornea has five distinct layers from anterior to posterior (Fig. 12.2).

1. *Epithelium* is a continuation of the epithelium of bulbar conjunctiva and consists of five layers of cells. The deepest layer has a palisade-like arrangement, the middle layers comprise polygonal cells and the superficial layer is formed by a flat squamous epithelium without keratinization.
2. *Bowman's membrane* is a thin structureless layer of about 12 μ thickness, placed between the epithelium and substantia propria. It is not a true elastic lamina; once destroyed, it does not regenerate. However, it shows a considerable resistance to infection.

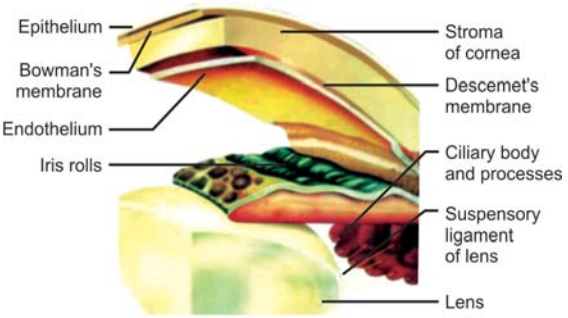


Fig. 12.1: Gross anatomy of cornea
[*Courtesy*: Allergan (India)]

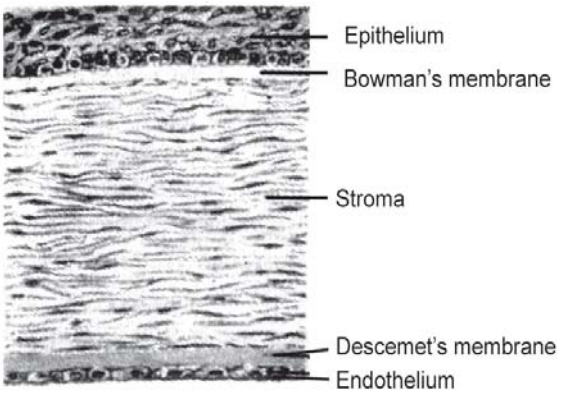


Fig. 12.2: Layers of cornea

3. *Substantia propria* or *corneal stroma* constitutes 90% of the entire thickness of the cornea and is composed of a modified connective tissue containing lamellae and cells. The lamellae, numbering 100 to 150, are ribbon-like bands of collagen fibers. They run parallel to each other and also to the surface of the cornea and become continuous with scleral lamellae at the limbus. Two types of corneal cells, keratocytes (fixed cells), and wandering cells are found between the lamellae.
4. *Descemet's membrane* is a strong, homogeneous structureless layer. It is sharply defined from the corneal stroma and quite resistant to the inflammatory process of the cornea. Even when the entire cornea gives way, the membrane remains unimpaired. Unlike Bowman's membrane, Descemet's membrane can regenerate. It ends abruptly at the limbus as the ring of Schwalbe. The posterior surface of the membrane may present wart like bodies—*Hassall-Henle bodies*.
5. *Endothelium* is the most posterior layer of the cornea and consists of a single layer of flat hexagonal cells. On slit-lamp biomicroscopy, they appear as a brown mosaic of polygons. The cell population of endothelium varies considerably in individual eyes and decreases with advancing age.

Blood Supply of the Cornea

The cornea is an avascular tissue. However, it does get some nourishment from the superficial plexus formed by the episcleral branches of the anterior ciliary arteries. The veins follow a similar course. The lymphatics are absent from the cornea.

Nerve Supply of the Cornea

The cornea is richly supplied by the ophthalmic division of the trigeminal nerve through the long ciliary nerves which come from the suprachoroidal space and enter the sclera. They pass into the cornea as 60 to 80 myelinated trunks after forming a pericorneal plexus. They divide into anterior and posterior groups, each comprising 40 to 50 twigs. The anterior group forms the subepithelial and intraepithelial plexuses, while the posterior supplies the posterior peripheral part of the cornea.

DISEASES OF THE CORNEA

Diseases of the cornea are serious as they often disturb the transparency of the cornea leading to visual impairment ranging from slight blurring to total blindness. The diseases of the cornea may be:

1. Inflammatory
2. Degenerative
3. Developmental, and
4. Symptomatic.

Inflammation of the Cornea

The inflammatory conditions of the cornea are frequent in occurrence and may arise from:

1. *Exogenous infection*: Cornea is often involved by direct invasion of an organism (corneal ulcer) which may or may not be preceded by trauma.
2. *Endogenous infection*: Cornea is involved in systemic, allergic and hypersensitivity reactions such as interstitial keratitis.
3. *Secondary infection*: Cornea is secondarily involved in the diseases of conjunctiva, sclera and uvea as these structures are in direct anatomical continuity with the cornea.

Classification

The inflammation of the cornea is known as *keratitis*. It may be of two types:

1. Ulcerative keratitis wherein the corneal epithelium shows discontinuity, and
2. Non-ulcerative keratitis wherein epithelium is intact.

Ulcerative Keratitis

Clinically, ulcerative keratitis is divided into two categories: superficial and deep.

Topographically, the ulcerative lesion (corneal ulcer) may be central, paracentral or marginal. The ulcer can be suppurative (purulent) or non-suppurative (nonpurulent).

Deep corneal ulcers may cause sloughing of the corneal stroma (sloughing corneal ulcer) or are associated with pus in the anterior chamber (hypopyon corneal ulcer). The deep progressive corneal ulcer may undergo perforation.

A simplified classification of ulcerative keratitis is given in Table 12.1.

Bacterial Corneal Ulcer

Bacterial corneal ulcer is an infection of the cornea associated with discontinuity of the corneal epithelium often accompanied with pain and diminution of vision.

Etiology Corneal ulcer occurs usually due to exogenous infection by pyogenic organisms, viz. *Staphylococcus aureus, Pseudomonas aeruginosa, Streptococcus pneumoniae, Neisseria gonorrhoeae* and *Streptococcus hemolyticus*. The ulcer is often associated with risk factors that disturb the integrity of the corneal epithelium. The common risk factors include trauma, foreign body, contact lens wear, prolonged use of corticosteroids and general disability or impaired defence mechanism. The intact corneal epithelium offers considerable resistance to the invasion by the microorganisms except *Neisseria gonorrhoeae* and *Corynebacterium diphtheriae*.

Pathogenesis The pathogenesis of corneal ulcer may be described under 4 stages.

Stage of infiltration: Corneal inflammation begins with local production of cytokines and chemokines inducing diapedesis and migration of neutrophils into the cornea from the limbal vessels. Some organisms produce proteases that disturb the extracellular matrix. The superficial layers of cornea show focal infiltration with inflammatory cells predominantly polymorphonuclear. The epithelium is edematous and raised at the site of infiltration. It undergoes necrosis and ultimately desquamates. If the lesion is superficial and does not involve Bowman's membrane it heals quickly without leaving any opacity. In case,the infiltration extends into the deeper layers of the cornea and destroys Bowman's membrane, it indicates the progression of lesion.

Stage of progression: The epithelium at the margins of the ulcer swells and overhangs. The corneal lamellae imbibe fluid and project above the surface giving a saucer-shaped appearance to the ulcer. The floor and the margin of the ulcer are packed with inflammatory cells and they appear gray. Enzymes released by neutrophils and activation of corneal metalloproteinases exacerbate necrosis. Bacterial toxins may diffuse in the anterior chamber and cause damage to the corneal endothelium, and induce iritis.

Table 12.1: Classification of ulcerative keratitis

Location	*Depth*	*Necrosis*	*Uveal reaction*	*Etiology*
Central Paracentral Peripheral	Superficial Deep	Suppurative Nonsuppurative	With hypopyon Without hypopyon	*Infective:* Bacterial, Viral, Fungal, Chamydial, Protozoal, Spirochetal *Allergic:* Phlyctenular, Vernal, Atopic *Traumatic* *Trophic* *Associated with systemic disorders* *Idiopathic*

Stage of regression: The sloughed corneal lamellae are cast off and the ulcer appears somewhat larger but clean with smooth floor and edges. Simultaneously, the limbal vessels extend and invade the ulcer. The vessels help in the proliferation of granulation tissue, supply of antibodies and sliding of marginal epithelium to bridge the gap.

Stage of cicatrization: In this stage, the granulation tissue is formed which is composed of irregularly arranged fibroblasts. Thus after healing, cornea becomes opaque at the site of ulceration. At times, due to deficient cicatrization, a corneal facet is left behind. The opacity in the cornea, depending on its density, may be nebular, macular or leukomatous.

Clinical features Pain, gritty sensation, redness, lacrimation, photophobia, blepharospasm and impairment of vision are the common symptoms of a corneal ulcer. Most corneal ulcers start as a gray or white localized infiltrate in the cornea causing loss of luster of the tissue. There is a discontinuity of the corneal surface which can be demonstrated by fluorescein staining. The ulcer takes a green stain (Fig. 12.3). The ulcer may be round, oval or irregular in shape.

The margin of the ulcer is edematous and overhanging with sloping edges. There occurs marked ciliary injection associated with slough (Fig. 12.4). Small superficial vessels grow in from the limbus towards the margin of the ulcer. Occasionally, an exuberant fibrofleshy growth may cover the ulcer and retard its healing.

Complications Generally, the ulcer heals by granulation. However, in adverse circumstances (like debility state or microorganism not amenable to the treatment), the ulcer extends both in size and depth. The corneal lamellae are macerated and the necrotic tissue covers the floor. The loss of entire corneal stroma leads to exposure of Descemet's membrane which may bulge as a transparent vesicle under the effect of normal intraocular pressure. The bulging of the Descemet's membrane is called *descemetocele* or *keratocele* (Fig. 12.5). The ulcer may eventually perforate if not managed properly.

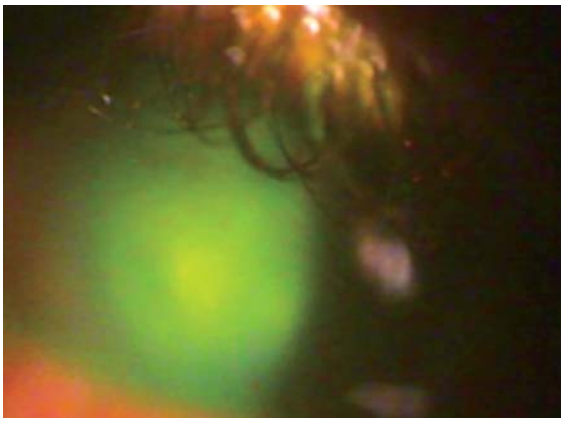


Fig. 12.3: Fluorescein staining of corneal ulcer

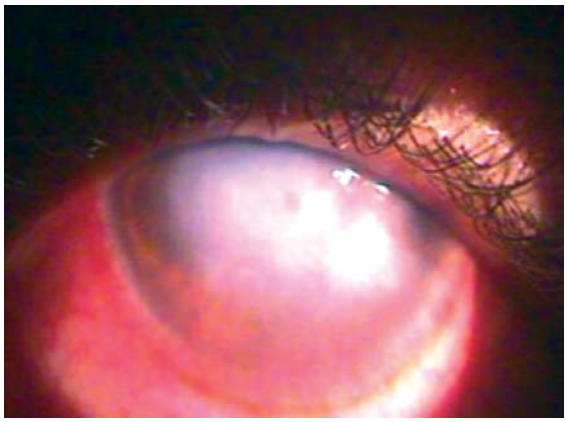


Fig. 12.4: Central corneal ulcer

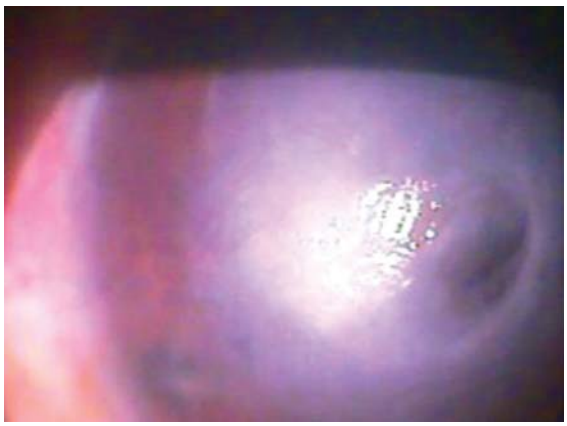


Fig. 12.5: Sloughing corneal ulcer with descemetocele

Specific Types of Bacterial Corneal Ulcers

Hypopyon Corneal Ulcer

A disk-shaped central corneal ulcer with hypopyon (sterile pus in the anterior chamber) and violent iridocyclitis is called *hypopyon corneal ulcer*.

Etiology The hypopyon ulcer is usually found in old, debilitated, malnourished patients who may be suffering from chronic dacryocystitis. There is always a risk of development of hypopyon ulcer following an injury by organic matters like leaf, twigs, coal, stone and finger-nail. *Streptococcus pneumoniae, Streptococcus hemolyticus, Neisseria gonorrhoeae* and *Proteus vulgaris* are common pyogenic organisms capable of producing the ulcer. *Pseudomonas pyocyanea* causes a fulminant sloughing hypopyon corneal ulcer with a greenish look within a short time. Hypopyon is frequently found in mycotic corneal ulcers.

Clinical features A typical pneumococcal ulcer, also known as *ulcus serpens*, starts as a grayish-white disk with infiltrating edges near the central part of the cornea. The cornea becomes cloudy and lusterless. The toxins liberated by the offending organisms diffuse into the anterior chamber and induce severe iridocyclitis associated with pouring of polymorphonuclear leukocytes in the anterior chamber known as *hypopyon* (Fig. 12.6A). The hypopyon remains sterile as long as Descemet's membrane is intact, since the latter is impermeable to the organisms. The hypopyon gravitates to the bottom of the anterior chamber (Fig. 12.6B). The horizontal upper level of the fluid moves with the change in the position of the patient's head. Hypopyon may be so small that the rim of sclera covers it and thus is hardly visible, or it may be so massive that it masks the entire iris. Large hypopyon tends to get organized owing to the presence of fibrinous network that traps the leukocytes.

The ulcer progresses on the edge of densest infiltration which appears as a yellowish crescent.

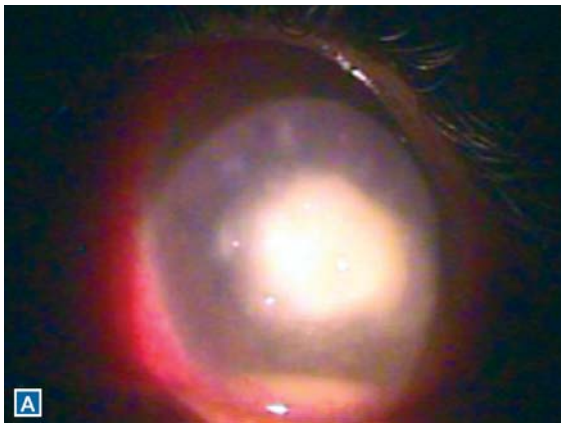


Fig. 12.6A: Hypopyon corneal ulcer

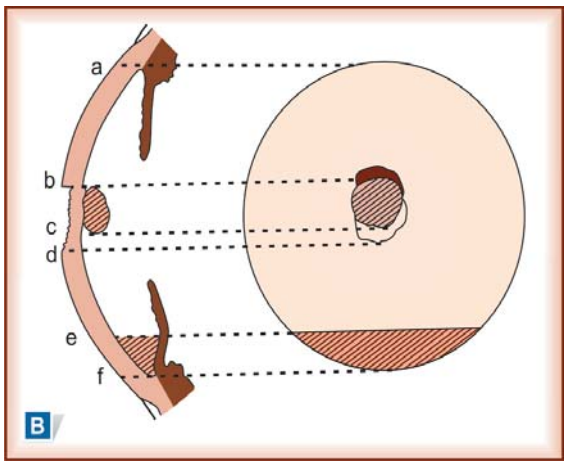


Fig. 12.6B: Schematic diagram of hypopyon corneal ulcer: b-d, Extent of ulcer; b, Actively progressive margin; b-c, Mass of leukocytes and fibrin adherent to the endothelial surface; e-f, Hypopyon

The superficial corneal stroma becomes necrotic and breaks down. An additional infiltration develops anterior to Descemet's membrane at a spot just opposite to the floor of ulcer, while the intervening corneal lamellae are healthy. The progression of the ulcer from both the sides causes gross corneal necrosis. Massive hypopyon often causes rise in intraocular pressure (secondary glaucoma).

Complications In severe cases, the entire cornea is affected by the ulcerative process. A sudden exertion (coughing or sneezing) results in *perforation of the ulcer* which is marked by escape of aqueous humor, reduction in intraocular pressure and forward displacement of the iris and the lens.

Complications of a perforated ulcer are serious and may endanger the eyeball. The effect of perforation largely depends on its size and position. A small perforation in the peripheral or paracentral zone of the cornea is promptly plugged by the iris, which on healing leads to *adherent leukoma*. When the perforation is large, *iris prolapse* occurs through the site of perforation (Figs 12.7A and B). When perforation occurs in the center of the cornea, the pupillary margin fails to seal the gap. The lens comes in contact with the cornea and exudates fill the gap. There occurs repeated formation and collapse of the anterior chamber and subsequently corneal fistula develops associated with *anterior capsular cataract*. Sudden perforation of a large corneal ulcer may even cause *extrusion of the lens from the eye, prolapse of the vitreous* and *intraocular hemorrhage*.

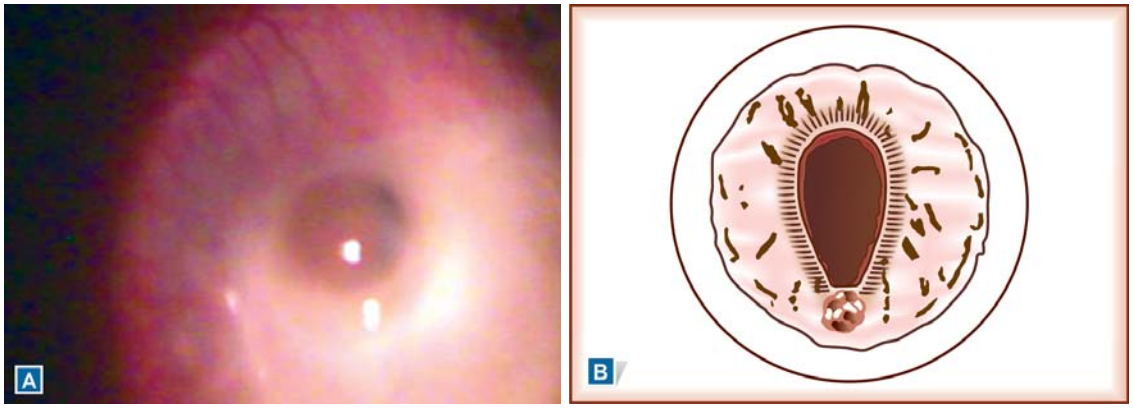
Sometimes the entire cornea sloughs off with the exception of a narrow rim at the margin causing a total prolapse of the iris. Such eyes go in *phthisis*. A pseudocornea is formed which ultimately tends to become ectatic. An anterior ectasia of the pseudocornea, in which the iris tissue is incarcerated, is called *anterior staphyloma*. The latter may be partial or total.

Staphylococcal Corneal Ulcer

Staphylococcal corneal ulcer is often found in compromised cornea, dry eyes and postherpetic keratitis. The ulcer remains localized with distinct borders and the surrounding stroma shows edema. Chronic ulcer tends to bore deep forming a stromal abscess which may cause perforation.

Pseudomonas Corneal Ulcer

Pseudomonas causes a rapidly spreading sloughing corneal ulcer with greenish-yellow mucopurulent discharge adherent to the ulcer (Fig. 12.8). The ulcer presents a characteristic diffuse epithelial graying that occurs away from the main site of epithelial and stromal infiltration. It spreads



Figs 12.7A and B: (A) Perforated corneal ulcer with prolapse of iris, (B) Diagrammatic representation of prolapse of the iris

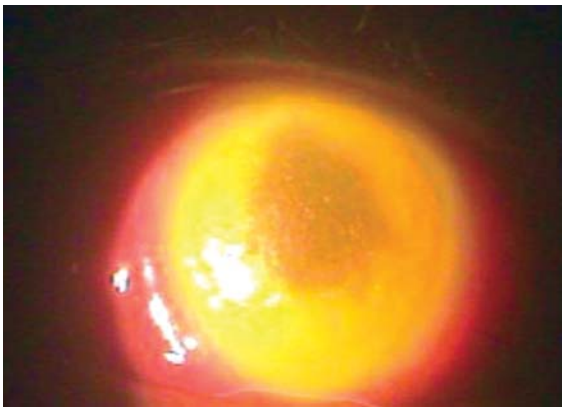


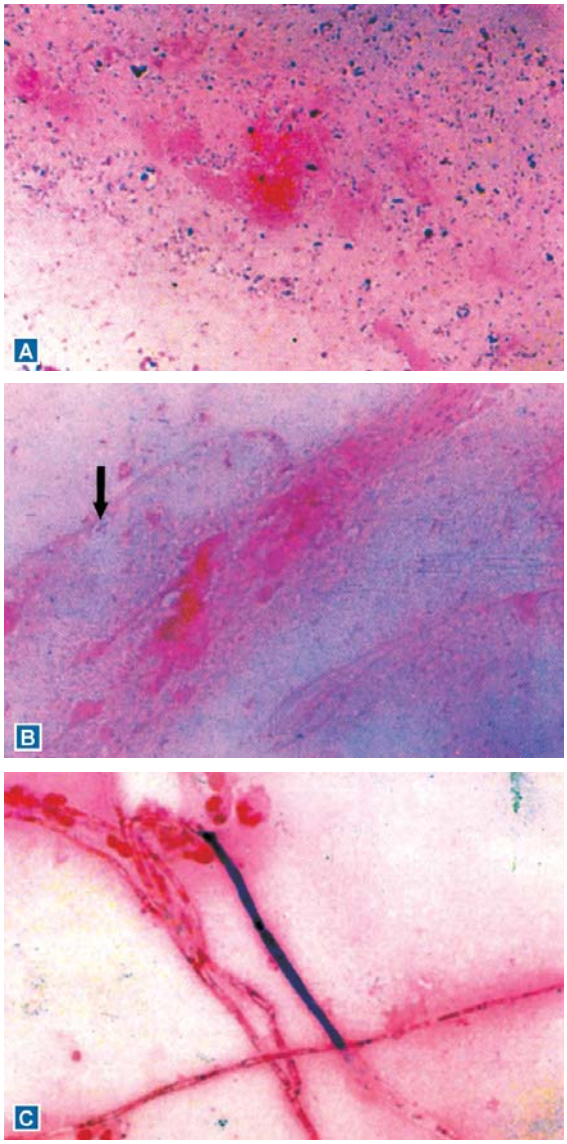
Fig. 12.8: Pseudomonas corneal ulcer

symmetrically and concentrically involving whole width and depth of the cornea associated with severe anterior chamber reaction and hypopyon formation. Pseudomonas strains produce protease, lipase and exotoxin-A that cause melting of the cornea resulting in perforation.

Moraxella Corneal Ulcer

Moraxella corneal ulcer occurs after trauma in diabetic or malnourished patients. The ulcer is usually oval in shape and located in the inferior half of the cornea. It is indolent and spreads deep into the stroma causing mild to moderate anterior chamber reaction.

Diagnosis A definitive diagnosis of corneal ulcer can only be made by organismal culture and sensitivity. The microbiologic work-up of an infective corneal ulcer must be done before the start of therapy. Smear and culture examinations should be carried out. The Kimura spatula or sterile disposable blade is used to take scrapings from the floor of the ulcer after anesthetizing the surface. The material is innoculated on blood agar, chocolate agar, and Sabouraud agar for culture and spread over slides for gram and Giemsa stains for bacteria, and KOH preparation for fungi. The



Figs 12.9A to C: Corneal scrapings stained by gram stain showing: (A) gram-positive cocci, (B) gram-negative bacteria, (C) septate fungal filaments (*Courtesy*: Dr Savitri Sharma, LVPEI, Hyderabad)

calcofluor white stained corneal scrapings are seen under fluorescent microscope for the fungal filaments (Figs 12.9A to C). Cultures are more sensitive than smears in identifying the causative organism.

Treatment

Prophylaxis: In majority of cases, development of corneal ulceration can be prevented by wearing protective glasses against foreign body and mechanical or chemical injuries, and proper and timely treatment of acute conjunctivitis, dacryocystitis and trichiasis. Exposure of the cornea should be prevented during unconsciousness or moribund conditions.

Once corneal ulcer develops, it requires prompt and adequate treatment. Surgical cleanliness, specific treatment of the infection, rest and protection to the eye are the basic principles of management of corneal ulcer.

Irrigation: Eye is irrigated with warm saline 2 to 3 times a day. It removes the discharge and necrotic materials along with organisms and their toxins. Warmth of the saline employed prevents vascular stasis and encourages flow of antibodies.

Antibiotics: The infection is controlled by the topical use of specific bactericidal or bacteriostatic antibiotics selected after the sensitivity test. However, testing facilities are not available in all the hospitals and the procedure is time consuming. It is, therefore, necessary to start a broad-spectrum antibiotic without waiting for the culture and sensitivity report. Instillations of fluoroquinolone (moxifloxacin, gatifloxacin or ciprofloxacin) and cefalosporin (cefazolin 5%) drops are effective in controlling the corneal infection caused by both gram-positive and gram-negative organisms.

Fortified antibiotics: In severe infection, fortified antibiotics are preferred to their commercially available concentrations. The fortified antibiotics used for the treatment of corneal ulcer are freshly prepared from their injectable preparations (Table 12.2).

To achieve therapeutic corneal concentration of the drug one of the antibiotics should be instilled every 5 or 10 minutes, then one drop every 30 or 60 minutes for 24 to 48 hours in day time. The antibiotic ointment can be applied at bed time.

Table 12.2: Fortified concentration of topical antibiotics and dosage of antibiotics for subconjunctival injection

Antibiotics	*Fortified concentration*	*Subconjunctival dose*
1. Amikacin sulphate	50 mg/ml	25 mg/0.5 ml
2. Cefaloridine	50 mg/ml	100 mg/0.5 ml
3. Cefamandole	50 mg/ml	100 mg/0.5 ml
4. Cefazolin	50 mg/ml	100 mg/0.75 ml
5. Ceftriaxone	50 mg/ml	100 mg/0.5 ml
6. Gentamicin	10-20 mg/ml	20 mg/0.5 ml
7. Tobramycin	10-20 mg/ml	20 mg/0.5 ml
8. Ciprofloxacin	3 mg/ml	—
9. Moxifloxacin	3.5 mg/ml	—
10. Gatfloxacin	3.5 mg/ml	—

The therapy should be reviewed or modified after receiving the culture-sensitivity report. The frequency of instillation of an antibiotic drop is gradually reduced if the condition improves.

Cycloplegics: Rest to the eye can be provided by the use of a cycloplegic, homatropine (2%) eye drop or atropine sulphate (1% drop or ointment) at least twice a day. Atropine prevents or relieves the ciliary spasm and minimizes the complications of accompanying anterior uveitis. Corticosteroid preparation must not be applied in a corneal ulcer (with rare exceptions) as it retards the epithelial healing and promotes secondary viral and fungal infections.

Protection to eye: The eye may be protected by pad and bandage unless there is a copious discharge. Eyes with discharge can be protected with dark glasses.

General measures: Systemic analgesics and anti-inflammatory agents (diclofenac sodium, ibuprofen, etc.) relieve pain from inflammatory reaction. General health of the patient must not be ignored. Malnutrition should be taken care of and diabetes, if present, be controlled.

Non-healing ulcer: In case, the ulcer does not respond favorably to the above therapeutic regimen and continues to progress, a thorough

search must be made for the presence of trichiasis, dacryocystitis, intracorneal foreign body and raised intraocular pressure. If found, corrective measures must be taken to hasten the healing.

Subconjunctival injections: Additionally, subconjunctival and intravenous (IV) injections of antibiotics are recommended as adjunctive therapy to topical fortified antibiotics (Table 12.2).

Specific indications for subconjunctival injection are: (i) non-healing ulcers with deep infiltrates, (ii) impending corneal perforation, and (iii) hypopyon corneal ulcer. Depending upon the culture-sensitivity test, subconjunctival injections of amikacin, gentamicin or cefalosporin may be given daily or on alternate day basis. However, injections are painful, anxiety provoking and may cause subconjunctival hemorrhage.

Surgical measures: To hasten healing, surgical measures may have to be adopted. These include: (i) mechanical debridement of the ulcer floor, (ii) cauterization of the ulcer floor with pure carbolic acid or 10% trichloracetic acid, and (iii) excision of 2 mm strip of limbal conjunctiva (peritomy) to regress exuberent corneal vascularization impeding healing.

Prevention of perforation: In case there is an imminent danger of corneal perforation, immediate measures should be taken to prevent perforation by lowering the intraocular pressure and by supporting the thin cornea. The intraocular pressure is reduced by oral acetazolamide and/or IV mannitol and topical aqueous suppressants. The procedure not only checks perforation, but also improves nutrition of the diseased cornea. Bandage contact lens, conjunctival flapping and tectonic corneal grafting can support the weak cornea.

Management of perforated corneal ulcer: In spite of these measures if cornea perforates, attempts should be made to restore the integrity of the anterior chamber as quickly as possible. Conjunctival flapping, penetrating therapeutic keratoplasty, cynoacrylate glue and collagen plug or shield may be helpful to save the eye.

Viral Corneal Ulcers

Both DNA and RNA viruses may cause eye diseases. However, DNA viruses such as herpes, vaccinia and adenovirus quite often infect the cornea. The corneal involvement in viral infections is described under the nonulcerative keratitis.

Fungal Corneal Ulcer

Etiology Fungal corneal ulcer occurs more frequently in warm and humid climate. Trauma to the cornea by vegetable material, indiscriminate use of corticosteroid, trauma related to contact lens wear, chronic keratitis and corneal surgery are important risk factors for fungal corneal ulcer. *Aspergillus fumigatus*, *Candida albicans*, *Fusarium*, and a few species of dermatophytes can cause fungal corneal ulcer.

Clinical Features Classically, most mycotic ulcers are gray, indolent and slowly progressive with relatively minimal symptoms. The clinical signs of keratomycosis include severe ocular reaction associated with ciliary injection, elevated rolled-

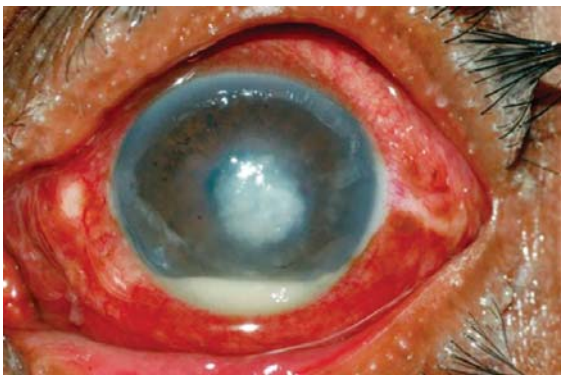


Fig. 12.10: Fungal corneal ulcer (*Courtesy*: Dr Lalitha Prajana, Aravind Eye Hospital, Madurai)

out hyphate margins, small round satellite lesions, dense white plaque on the corneal endothelium and non-sterile hypopyon (Fig. 12.10). Sometimes a white immune ring (*Wesseley ring*) is seen in the mid-periphery of the cornea.

The mycelia of the causative fungus can be demonstrated in scrapings after fixation with KOH.

Treatment Natamycin (5%) suspension is used to manage corneal ulcer caused by *Fusarium, Candida albicans* and filamentous fungi. Topical amphotericin B (1-10 mg/ml) eye drop is effective against *Aspergillus* and *Candida*. Fluconazole (0.2%) ophthalmic solution is active against *Candida albicans.* Nystatin and miconazole (2%) eye ointments can be used to manage fungal corneal ulcer. Oral antifungal agents are used when there is suspicion of endophthalmitis.

Marginal Corneal Ulcer (Catarrhal Ulcer)

Etiology Marginal corneal ulcers are frequently seen in old people and thought to be caused by *Staphylococcus aureus*, *Morax-Axenfeld diplobacilli* or *Haemophilus aegyptius*. They are often associated with chronic blepharoconjunctivitis. They are thought to be caused by hypersensitivity reaction to the exotoxins of *Staphylococcus aureus*.

Clinical features Ocular discomfort, mild pain, watering and photophobia are presenting symptoms. The ulcer appears as shallow infiltrated crescent at the corneal periphery with vascularization, and runs a chronic indolent course but seldom perforates. Occasionally, a deep marginal gutter may develop which covers the entire periphery of the cornea forming a ring ulcer. It can lead to necrosis of the cornea.

Treatment Oxytetracycline is very effective in the management of Morax-Axenfeld infection. Atropine drops or ointment should be used to prevent complications. Topical instillations of antibiotic-corticosteroid drops provide quick relief. Ciprofloxacin and norfloxacin are effective in Haemophilus infection.

Idiopathic Corneal Ulcer or Mooren's Ulcer (Chronic Serpiginous Ulcer)

Mooren's ulcer is a chronic, progressive, painful, idiopathic ulceration of the peripheral cornea.

Etiology The etiology of Mooren's ulcer is unknown. There is a growing evidence to suggest that autoimmunity plays a key role in its causation. Accidental trauma, surgery and helminthic infestation are considered as risk factors. Mooren's ulcer is more common in African countries where parasitic infestations are endemic.

Clinical features

Mooren's ulcer occurs in old age. The ulcer may cause pain, lacrimation, photophobia and impairment of vision. The ulcer starts in the periphery of the cornea and slowly spreads cirumferencially and centripetally (Fig. 12.11). Typically its advancing edge undermines the corneal epithelium and superficial stroma and destroys Bowman's membrane. The ulcer progresses in the direction of its advancing edge, while cicatrization starts at the periphery. Minor trauma

Fig. 12.11: Mooren's ulcer

and secondary bacterial infection may lead to perforation. Extensive vascularization and scarring of the cornea may develop.

Treatment Currently there is a lack of effective treatment for Mooren's ulcer. Besides topical cycloplegic and antibiotic, topical corticosteroids, acetylcysteine (10%), and topical cyclosporine are used with variable success. Administration of systemic corticosteroids, methotrexate, cyclophosphamide or cyclosporine has given encouraging results. Use of contact lens, conjunctival excision and lamellar keratoplasty have been recommended.

Atheromatous Corneal Ulcer

Atheromatous corneal ulcer generally occurs in an old leukoma undergoing degenerative changes with calcareous deposits. As the cornea is devitalized and insensitive, it is readily vulnerable to infection. The ulcer perforates quickly and panophthalmitis supervenes. It should be treated on the general lines of a corneal ulcer but if the eye is blind, evisceration relieves the patient of unnecessary agony.

Corneal Ulcer Associated with Malnutrition

Malnutrition lowers the general body resistance and predisposes the individual to infection. Following two types of corneal ulcers are found in poorly nourished children: central marasmic ulcer and keratomalacia.

Central Marasmic Corneal Ulcer

Bilateral, central, superficial or deep corneal ulcer with vascularization may occur in marasmic children. It is prone to rapid perforation. Routine treatment of corneal ulcer along with improvement of the general health of the child helps in healing.

Keratomalacia

Bilateral melting of the cornea associated with xerosis of the conjunctiva and vitamin A deficiency characterize keratomalacia.

Etiology Keratomalacia results from vitamin A deficiency either due to its poor dietary intake or impaired absorption from the gastrointestinal tract.

Clinical features Keratomalacia is often seen in children below 5 years of age belonging to underprivileged families. They look extremely ill and suffer from infective diarrhea. Night-blindness and conjunctival signs of vitamin A deficiency precede keratomalacia. Cornea becomes dull and insensitive and a keratinized plaque may cover the surface. The ulcer is round or oval with dense yellowish-white infiltration near the central part of the cornea. Typically, it is devoid of usual inflammatory reaction. The lesion is rapidly progressive and involves full-thickness of the cornea. Soon the necrotic stroma sloughs off causing perforation, extrusion of the intraocular contents and loss of eyeball.

Treatment The treatment must be aimed to improve the general health of the child. Vitamin A capsule 200000 IU should be administered in 3 doses as recommended by the WHO. Injections of vitamin A in aqueous (50000 to 100000 IU) can be given weekly. The infective diarrhea must be controlled by suitable drugs. Electrolyte and fluid imbalance should be corrected by intravenous drip. Atropine and broad spectrum antibiotics are locally applied and eye is protected by a bandage or dark glasses. Prompt and energetic treatment can save the eye from blindness.

Nonulcerative Keratitis

Nonulcerative keratitis is divided into two broad groups: superficial and deep.

Superficial Keratitis

Superficial keratitis involves the corneal epithelium, Bowman's membrane and superficial corneal stroma. It can be further divided into following categories:

1. Superficial punctate keratitis
2. Thygeson's superficial punctate keratitis
3. Superficial limbic keratitis
4. Filamentary keratitis
5. Corneal erosions.

Superficial Punctate Keratitis

Multiple, small, pin-head size round lesions in the epithelium and superficial stroma of the cornea characterize superficial punctate keratitis (SPK).

Etiology SPK is commonly seen in viral infections of the eye (adenovirus, herpes simplex and herpes zoster), chlamydial infection, staphylococcal blepharoconjunctivitis, dry eye syndrome and following trauma to the eye.

Clinical features Photophobia, watering and mild ocular discomfort are common presenting symptoms. Vision may or may not be affected. Punctate epithelial lesions stain with fluorescein but the subepithelial lesions do not. SPK secondary to staphylococcal conjunctivitis involves the lower-third of the cornea while trachoma affects the upper-third.

Treatment Generally, SPK responds to topical corticosteroid therapy. However, corticosteroids must not be used in herpetic infections. Here the use of acyclovir is beneficial. Frequent instillations of tear substitutes give relief in dry eye syndrome.

Thygeson's Superficial Punctate Keratitis

Thygeson's superficial punctate keratitis is a non-contagious, bilateral, asymmetrical, coarse epithelial keratopathy unassociated with conjunctival injection.

Etiology The disease is of unknown etiology. However, viral infection is implicated. The rapid response to corticosteroids suggests that it is an immunemediated disease.

Clinical features Foreign body sensation in the eyes, watering and photophobia are usual complaints of the patient. Although the conjunctiva seems normal, there are multiple coarse, round or oval, slightly elevated flecks of opacity in the cornea. Corneal lesions stain with fluorescein and rose bengal dyes. The disease has a chronic course. Exacerbation and remissions are common.

Treatment Mild cases may not need any treatment except artificial tears. Topical steroid, fluorometholone 0.1% drops 4 times a day, gives relief during exacerbation but it should be gradually tapered as soon as the corneal epithelial stain becomes negative. Bandage contact lens may be needed in resistant cases.

Superior Limbic Keratoconjunctivitis

Marked injection of the superior limbus and the upper palpebral conjunctiva and the presence of punctate keratitis in the upper half of the cornea characterize superior limbic keratoconjunctivitis (SLK).

Etiology Superior limbic keratoconjunctivitis has an obscure etiology. The disease is predominantly seen in females and in patients with hypothyroidism.

Clinical features Ocular discomfort, irritation and mild lacrimation are common presenting symptoms. The disease involves both eyes. Papillary hyperplasia of the upper palpebral and limbic conjunctiva is marked. Superficial punctate lesions in the upper part of the cornea often stain with fluorescein and rose bengal.

Treatment Artificial tears and corticosteroid drops may provide transient relief. Bandage contact lens,

diathermy of the upper limbus or peritomy may be helpful for the management of SLK.

Filamentary Keratitis

Filamentary keratitis is characterized by the presence of mucous filaments associated with superficial keratitis.

Etiology It is found in patients with kerato-conjunctivitis sicca, recurrent corneal erosions, neurotrophic keratopathy, herpes simplex keratitis and prolonged eye patching.

Clinical features The filament comprises a mucous core surrounded by the corneal epithelium. The one end of the filament is attached to the epithelium while the other moves freely over the cornea. The closure of the lids or eye movements put tension on the filaments causing pain and foreign body sensation.

Treatment The condition is treated by instillation of hypertonic saline (5%) and manual removal of filaments and short-term patching of the eye. The use of topical 10-20 % acetyl cysteine benefits the patient as it is a mucolytic agent. Bandage contact lens is also helpful.

Corneal Erosions

Etiology Punctate epithelial erosions of the cornea which stain with fluorescein are found in acute blepharoconjunctivitis. They are non-specific lesions and may be produced by toxins of staphylococci and viruses or by chemical irritants. The condition may be familial or associated with trauma or corneal dystrophy. The corneal erosion is a serious disorder and occurs due to a defect in the basement membrane of the corneal epithelium. The epithelial layers are loose and prone to separation and frequent erosions.

Clinical features Recurrent corneal erosions cause intense pain, photophobia, irritation and redness. Recurrent erosions often occur in the lower part of the cornea.

Treatment The erosions of the cornea should be treated as simple abrasions but recurrent erosions need careful management. Mechanical denuding of the corneal epithelium or cauterization may be necessary. The use of a bland ointment or hypertonic saline drops (5%) or ointment (6%) with pressure bandage may promote healing. X-ray therapy is said to be beneficial. Lamellar keratoplasty or bandage contact lens is helpful in indolent cases.

Photophthalmia

Etiology Photophthalmia may be caused by exposure to short wavelength ultraviolet rays either reflected from snow surface (snow-blindness) or from other sources (welding or short-circuiting of high-tension electric current). The essential pathology is the desquamation of corneal epithelium causing multiple erosions.

Clinical features Photophthalmia is characterized by photophobia, blepharospasm, burning and watering. The symptoms appear after a latent period of 5 to 6 hours of exposure. The corneal epithelium shows a breach in continuity that takes up fluorescein stain.

Treatment Prophylactic wearing of dark glasses is advisable. Once the condition develops, antibiotic ointment, cycloplegic drop and semi-pressure bandage (for a day) are recommended.

Viral Keratitis

Herpes Simplex Keratitis

The word 'herpes' is derived from a Greek word meaning 'to creep' or 'to move like a serpent'. In fact, it is a misnomer since the disease spreads by direct contact between two persons (kissing or sexual contact).

Etiology The disease is caused by herpes simplex virus (HSV). The two strains identified are HSV-1

and HSV-2. The former affects the upper part of the body, mostly the mouth, lips and eyes while the latter attacks essentially the ano-genital region. The infection is quite common, 50-90% of all human beings may suffer from herpes during their life-time. A person once infected becomes a carrier. An attack does not produce lasting immunity as recurrences are frequent, particularly associated with upper respiratory tract infection. The HSV infection occurs in two forms: primary and recurrent.

Primary Herpetic Infection

Primary herpetic infection is found in non-immune subjects. A high transference rate of HSV infection occurs in children under 3 years of age owing to close contact. The incidence of herpetic infection increases with age. Bilateral ocular involvement is seen in nearly 12% of cases. The infection remains subclinical in approximately 50% of the afflicted patients.

Clinical features The primary infection may take a mild or a fatal course if encephalitis develops. The disease may cause mild fever, malaise and regional lymphadenopathy. When the face is involved, skin lesions consisting of vesicles develop on the lips and periorbita. Blepharoconjunctivitis or acute follicular conjunctivitis associated with a watery discharge is not uncommon. A transient epithelial punctate keratitis may be found in nearly 50% cases of herpetic blepharoconjunctivitis. However, a coarse epithelial punctate keratitis may be the forerunner of a typical herpetic corneal lesion—dendritic keratitis (Fig. 12.12). Subsequently, subepithelial keratitis develops but disciform keratitis is a rarity. The primary herpetic infection is usually self-limiting, majority of the lesions heal without sequelae.

Recurrent Infection

During the primary infection herpes virus reaches the trigeminal ganglion where it may lie dormant for many years. Reactivation of the virus occurs following poor general body resistance (debilitating diseases, stress, trivial trauma, attack of flu, use of corticosteroids and other immunosuppressive agents). The virus travels down the trigeminal nerve to involve the eye. The recurrent infection is not associated with systemic features.

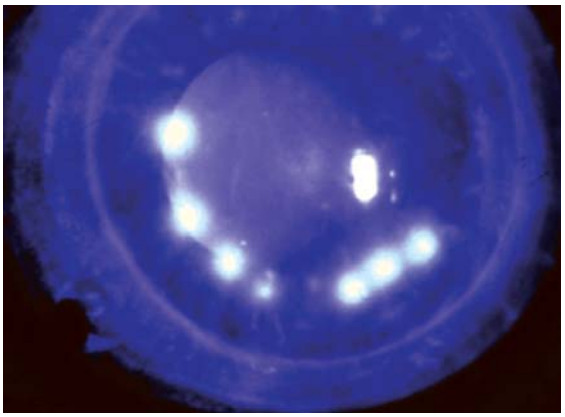
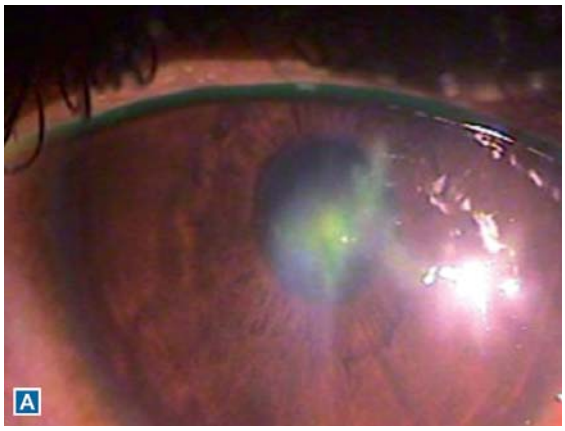


Fig. 12.12: Herpetic keratitis

Clinical features The clinical features of recurrent herpetic infection of the cornea vary largely. It may start with minimal symptoms of a foreign body sensation, watering and mild photophobia. The accompanied corneal hypoesthesia often causes the patient to delay the medical consultation. The corneal lesions can be epithelial or stromal.

Epithelial lesions: Foreign body sensation, discomfort in light, redness and blurred vision are common symptoms.The most characteristic lesion of herpes simplex recurrent infection is the *dendritic ulcer* (Figs 12.13A and B). It is caused by the multiplication of virus without cellular infiltration. The lesion is composed of clear vesicles in the epithelium arranged in a dendritic or stellate pattern. The terminal ends of dendritic figures are usually knobbed. Desquamation gives a linear branching ulcer which stains with fluorescein, while virus laden cells at the margin take rose bengal stain. Corneal sensitivity is usually diminished. The dendritic ulcer can



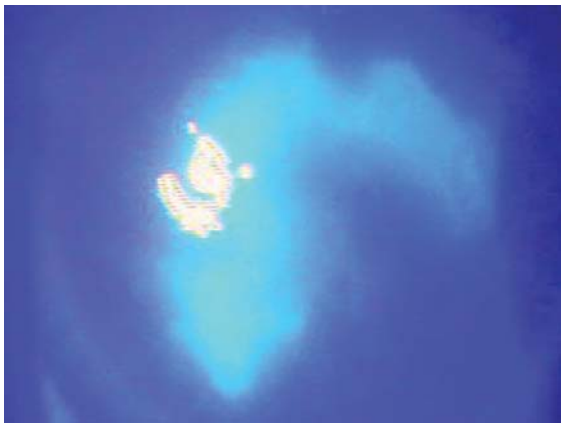
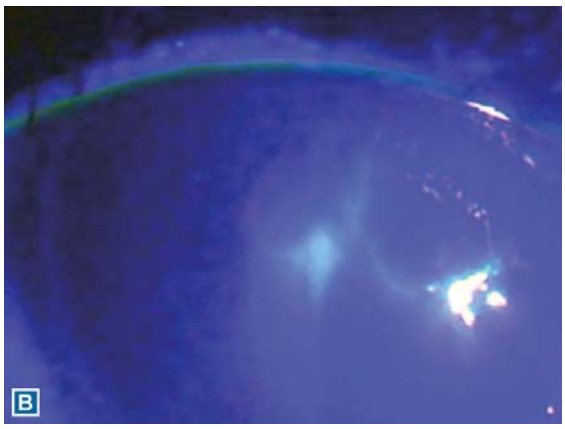


Fig. 12.14: Geographical ulcer



Figs 12.13A and B: Dendritic corneal ulcer: (A) Fluorescein stained, (B) Seen under cobalt-blue filter

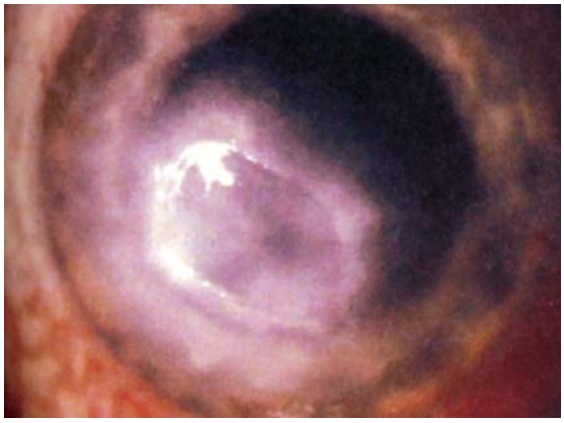


Fig. 12.15: Metaherpetic corneal ulcer (*Courtesy*: Dr AB Tullo, Manchester Royal Eye Hospital, Manchester)

spread in many directions and lead to an amoeboid configuration commonly known as *geographical ulcer* (Fig. 12.14). The geographical lesion is a result of rapid viral replication owing to reduced tissue resistance particularly after the indiscriminate use of topical corticosteroids.

Metaherpetic lesions: Recurrent corneal erosions in herpetic infection are not uncommon. The lesions are referred to as *trophic* or *metaherpetic keratitis* (Fig. 12.15). They are not caused by reactivation of the virus, but represent a persistent defect in the basement membrane. The margins of erosions do not stain with rose bengal.

Stromal lesions: Following several episodes of dendritic keratitis, stromal involvement usually occurs. Stromal lesions are mainly of two types—disciform keratitis and stromal necrotic keratitis.

Disciform keratitis or *nonnecrotizing stromal keratitis* or *immune stromal keratitis* is perhaps a hypersensitivity reaction of stroma to herpes infection. Marked impairment of vision, mild discomfort and watering are common symptoms. Disciform karatitis is characterized by a more or less central disciform edema of the cornea involving stroma as well as epithelium (Fig. 12.16). Stromal infiltrates are seldom seen, but a ring of infiltrates (*Wessely ring*) may be present. The stromal edema may be due to a low-

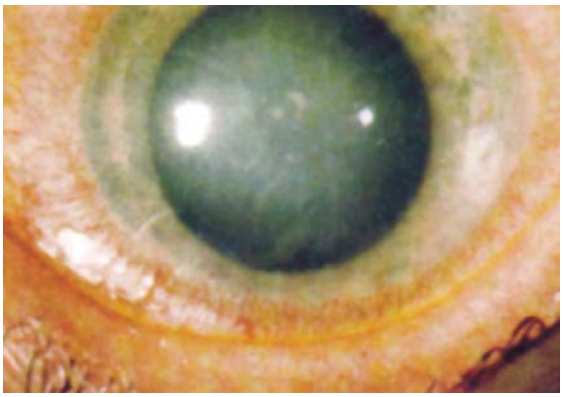


Fig. 12.16: Disciform keratitis (*Courtesy*: Dr AB Tullo, Manchester Royal Eye Hospital, Manchester)

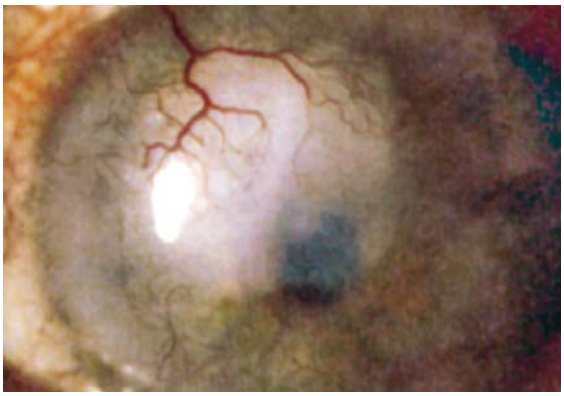


Fig. 12.17: Stromal keratitis with vascularization (*Courtesy*: Dr AB Tullo, Manchester Royal Eye Hospital, Manchester)

grade inflammation and damage to the underlying endothelium (*endothelitis)*. Other features of the disciform keratitis are folds in Descemet's membrane and a mild anterior uveitis with lymphocytic keratic precipitates. The presence of keratic precipitates and reduced corneal sensation is helpful in differentiating herpetic disciform keratitis from corneal hydrops.

Stromal necrotizing keratitis is an uncommon lesion caused by active invasion and destruction of corneal stroma by herpes virus. A typical lesion has a cheesy yellowish-white necrotic appearance similar to bacterial keratitis. The ciliary congestion is marked. It is associated with anterior uveitis and even spontaneous hyphema.

Complications Herpes simplex keratitis may progress and cause vascularization (Fig. 12.17), thick corneal scarring, descemetocele formation (Fig. 12.18) and perforation of the cornea (Fig. 12.19).

Diagnosis

The laboratory diagnosis of herpetic keratitis includes demonstration of multinucleated giant cells, intranuclear inclusions, and presence of HSV-1 antigen in corneal scrapings (Figs 12.20A and B). Polymerase chain reaction is a sensitive test for the diagnosis of herpetic infection.

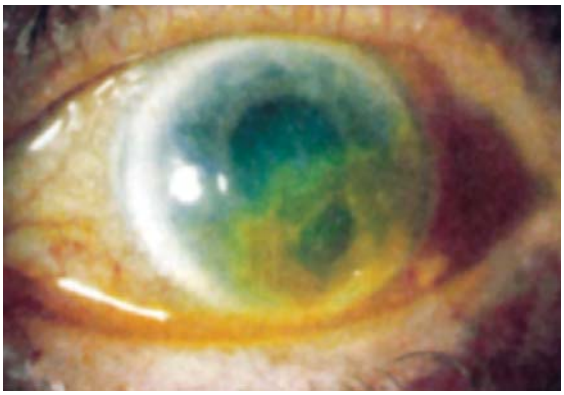


Fig. 12.18: Descemetocele following herpetic ulcer (*Courtesy*: Dr AB Tullo, Manchester Royal Eye Hospital, Manchester)

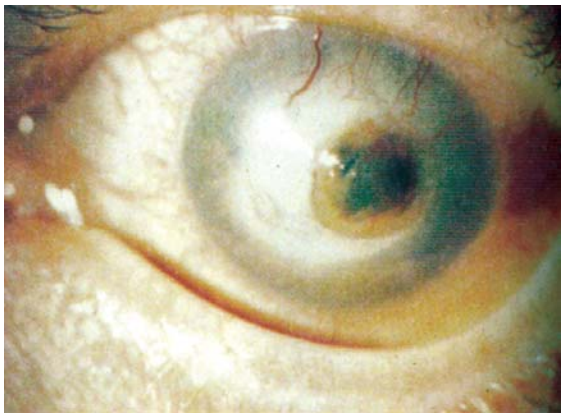
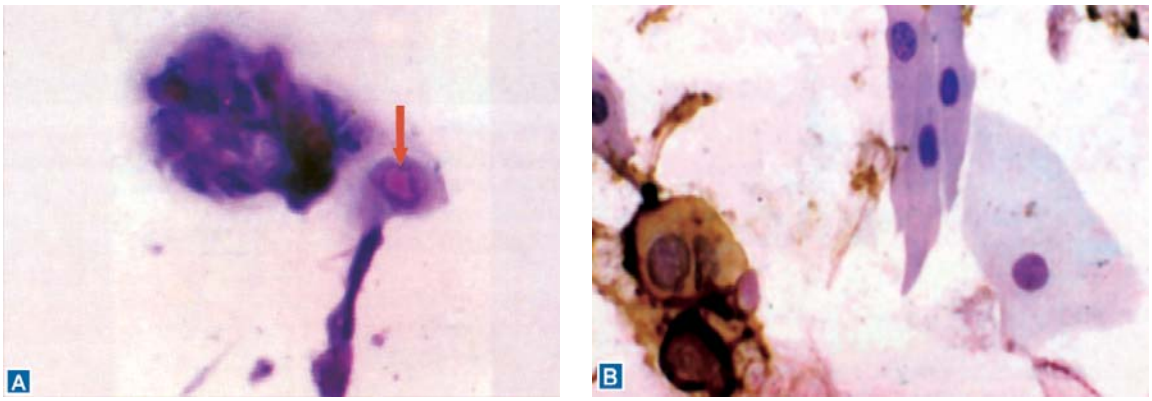


Fig. 12.19: Corneal perforation (*Courtesy*: Dr. AB Tullo, Manchester Royal Eye Hospital, Manchester)



Figs 12.20A and B: (A) Intranuclear inclusions, (B) HSV-1 antigen in epithelial cells stained brown (*Courtesy*: Dr Savitri Sharma, LVPEI, Hyderabad)

Treatment

Epithelial lesions: The primary herpetic infection is self-limiting and responds well to antiviral therapy. However, recurrent infections, particularly the stromal, pose serious therapeutic problem. A number of antiviral drugs such as 5-iodo-2-deoxyuridine (IDU), vidarabine (Vira-A) ointment, and trifluorothymidine (TFT) are effective in the management of epithelial lesions caused by HSV. Earlier 5-iodo-2-deoxyuridine (0.1% drops or 0.5% ointment) was used several times a day. The drug may cause corneal toxicity. Vidarabine 3% ointment 5 times a day and trifluorothymidine 1% drops 9 times a day are quite effective and less toxic.

Acycloguanosine (acyclovir) is a potent antiviral agent which can be used topically as well as orally. The 3% ointment form of the drug is applied 5 times daily for 2 weeks. Recent studies have shown that acyclovir-resistant strains of herpes simplex can be effectively treated by ganciclovir gel (0.15%) adopting similar treatment regimen. Resistant cases or recurrent infections are managed by debridement of corneal epithelium and a combination of topical and oral acyclovir (800 mg 5 times a day for 10-14 days). Besides the antiviral agents, topical cycloplegic drugs are always recommended for the management of HSK.

Metaherpetic lesions: Antiviral therapy is not needed in the management of metaherpetic keratitis. The erosions may heal with the use of artificial tear drops several times in a day and bandage soft contact lens.

Stromal lesions: Topical corticosteroids given together with acyclovir check the progression of HSV stromal keratitis (according to Herpetic Eye Disease Study). Topical 1% prednisolone may be used two hourly and gradually tapered. 3% acyclovir is used topically 4-5 times a day. Topically applied antiviral drugs are not absorbed by the cornea through intact epithelium; but orally administered acyclovir penetrates the intact cornea epithelium and anterior chamber. Therefore, oral acyclovir (800 mg 5 times a day for 2-3 weeks) is preferred in disciform keratitis and necrotizing herpetic stromal keratitis. However, the role of oral acyclovir in preventing recurrences is questionable.

Tectonic penetrating keratoplasty (PKP) is indicated in impending corneal perforation. Patients with stromal scarring may need PKP for visual improvement.

Herpes Zoster Ophthalmicus

Herpes zoster ophthalmicus (HZO) is characterized by eruption of multiple vesicles strictly on one side of the face along the distribution of ophthalmic division of the trigeminal nerve (Fig. 12.21), preceded by severe neuralgia and constitutional symptoms.

Etiology The disease is caused by varicella zoster virus. It is believed that the virus remains dormant after infection with chickenpox in young age and gets activated at a later stage causing herpes zoster ophthalmicus.

The essential lesion in herpes zoster ophthalmicus is an acute hemorrhagic necrotizing gasserian ganglionitis. It always involves the supraorbital, supratrochlear and infratrochlear branches and frequently the nasal branch of trigeminal nerve. Varicella zoster virus lies latent in sensory neural ganglion following the primary infection. An endogenous reactivation of latent virus occurs in elderly persons without any predisposing cause. However, the occurrence of HZO is more common in patients with acquired immune deficiency syndrome, malignancy or after exposure to radiation and debilitating disease.

Clinical features Ocular involvement occurs in more than 70% of patients with HZO affecting the first division of trigeminal nerve but it can also occur with the involvement of second division.

The disease starts abruptly with severe neuralgic pain along the distribution of the first division of trigeminal nerve often associated with fever, nausea, vomiting and malaise. The symptoms, especially the pain, diminishes within two to three days after the appearance of crops of vesicles on one side of the forehead and the scalp. The vesicles may also involve the nose as well as the lids and the cornea. The lids are edematous and the corneal lesion may vary from a small infiltrate to a big vesicle which may ulcerate.The eye is almost always affected if the vesicles appear on the tip and side of the nose supplied by the nasociliary nerve (*Hutchinson's rule*).

Mild cases of HZO may develop conjunctivitis, superficial punctate keratitis, subepithelial infiltrates, nummular keratitis and single or multiple microdendrites. The dendrites of herpes zoster are smaller without central ulceration and terminal bulbs, while that of herpes simplex have central ulceration and terminal bulbs.

Severe cases of HZO may present with stromal corneal lesions often associated with anterior uveitis (Fig. 12.22). A central disciform keratitis is

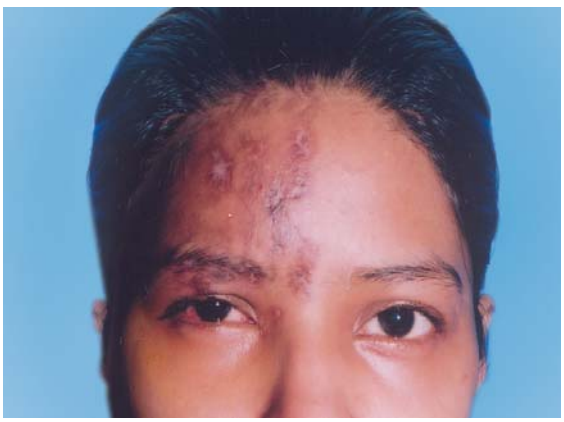


Fig. 12.21: Herpes zoster ophthalmicus

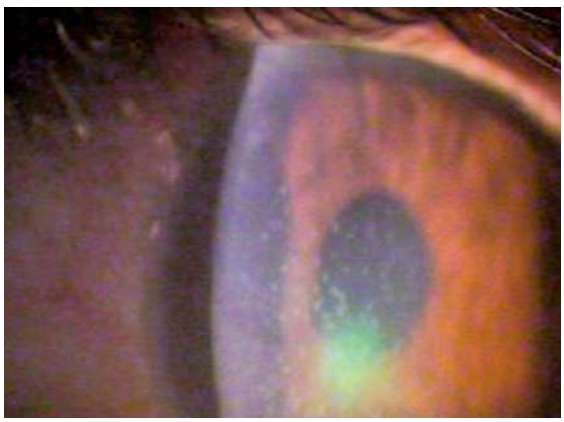


Fig. 12.22: Herpes zoster keratouveitis

usually preceded by nummular keratitis—multiple minute subepithelial deposits often associated with stromal haze.

Complications The skin vesicles are initially filled with a clear fluid but soon suppurate. Later, they form crusts and leave behind pitted scars in about three weeks' period. The affected area including the cornea is insensitive. Later, neurotrophic keratopathy, a very serious complication of the disease, may develop. Scleritis and iridocyclitis with multiple small keratic precipitates are not uncommon. HZO may sometimes lead to secondary glaucoma. However, in the early stage of the disease the ocular tension may be low. Sector iris atrophy, focal choroiditis, occlusive retinal vasculitis, anterior segment ischemia and retinal detachment may develop. Optic neuritis, extraocular muscle palsy and Bell's palsy are other complications of the disease. Rarely encephalitis may supervene. The patient may suffer from moderate to severe postherpetic neuralgia which persists for months to years.

Treatment Antiviral therapy should be started within 72 hours of the onset of skin lesions of HZO in order to decrease the incidence of postherpetic neuralgia and uveitis. Currently oral famciclovir 500 mg 3 times a day, valacyclovir 1 g 3 times a day or acyclovir 800 mg 5 times a day for 7 to 10 days are recommended. Intravenous acyclovir should be preferred in immunocompromised patients. Administration of oral corticosteroids reduces pain, prevents massive crust formation and facilitates early recovery. Topical antivirals are not effective. Topical cycloplegic and corticosteroids control keratouveitis. Neurotrophic keratopathy is managed with tarsorrhaphy.

Cutaneous lesions are treated with antibiotic-corticosteroid cream. Acyclovir skin ointment may be applied. Capsaicin cream may reduce postherpetic neuralgia. Amitriptyline or carbamazepine may be needed to control severe pain.

Vaccinia Keratitis

Accidental corneal involvement with vaccinia virus may be caused by autoinoculation from an arm pustule.

Vaccinia can cause superficial dendritic or geographical ulceration or a severe keratitis.

Topical and systemic vaccinia immunoglobulins may help in the resolution of lesion. There is some evidence to suggest that vidarabine monohydrate and interferon may accelerate healing.

Protozoal Keratitis

Acanthamoeba Keratitis

Acanthamoeba keratitis is an uncommon protozoal corneal infection characterized by severe pain, photophobia, lacrimation, blurred vision and a ring-shaped infiltrate surrounding a central corneal ulcer (Fig. 12.23). It is often found in contact lens wearers.

Etiology The keratitis is caused by a small protozoan, *Acanthamoeba*. The organism can adhere to the contact lens surface or may be present in nonsterile contact lens solution. Trauma may be a predisposing event.

Clinical features Severe ocular pain, photophobia, foggy vision and watering are common symptoms. Early lesion of Acanthamoeba keratitis

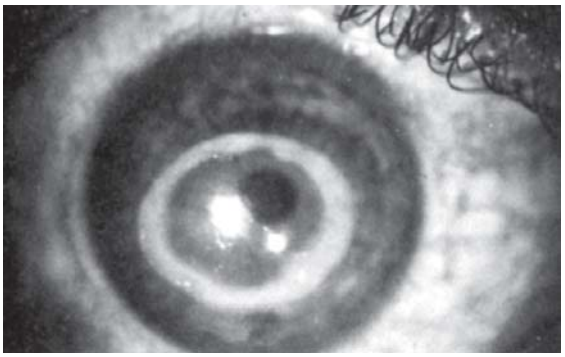


Fig. 12.23: Acanthamoeba keratitis
(*Courtesy*: LV Prasad Eye Institute, Hyderabad)

manifests as granular epithelial irregularities with punctate or dendritiform perineural infiltrates mimicking herpes simplex keratitis. Epithelial edema and erosions may occur. Patchy infiltrates develop in the anterior corneal stroma which coalesce and form an incomplete or a complete ring. Enlarged corneal nerves, called *radial perineuritis*, limbitis and nodular or diffuse scleritis may develop. Later, suppurative ulceration of the cornea or stromal abscess associated with anterior uveitis and hypopyon may supervene. Perforation is not rare. In spite of severe inflammation, corneal neovascularization is typically absent.

A history of soft contact lens wear or swimming in contaminated water, the characteristic clinical picture and demonstration of Acanthamoeba cysts on direct examination of corneal scrapings (Fig. 12.24) or on biopsy are diagnostic.

Treatment Proper cleaning and disinfection of contact lenses can prevent the occurrence of Acanthamoeba keratitis in contact lens wearers. Hydrogen peroxide and chlorhexidine solution can eradicate the organism from the contact lens.

Besides cycloplegic eye drops, treatment includes polyhexamethyl biguanide (PHMB) 0.02% or chlorhexidine 0.02% eye drop hourly for 1 week, then taper over 2-3 months, propamidine isethionate (0.1%) solution every 30 minutes to 2 hourly interval and dibromopropamidine (0.15%) ointment at night. Topical neomycin, paromomycin, polymyxin B, miconazole 1% and clotrimazole 1% are quite effective.

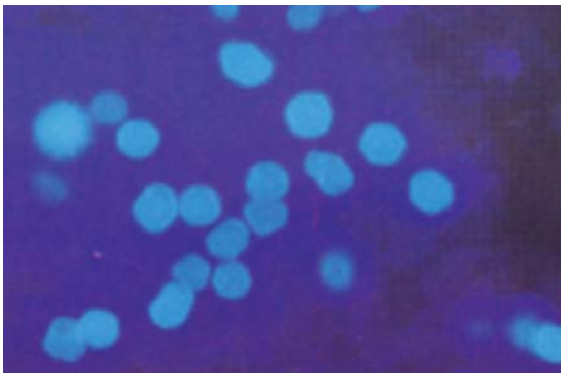


Fig. 12.24: Corneal scraping stained with KOH and calcofluor showing acanthamoeba cysts under fluorescent microscope (*Courtesy*: Dr Savitri Sharma, LVPEI, Hyderabad)

Traumatic Keratitis

Traumatic keratitis is described in the chapter on *Injuries to the Eye*.

Keratitis Secondary to Diseases of Conjunctiva

Phlyctenular Keratitis

Cornea is often involved in phlyctenular keratoconjunctivitis. Sometimes, phlyctens are located on the cornea and appear as gray nodules slightly raised above the corneal surface. The overlying epithelium breaks down and yellowish ulcers are formed.

Occasionally, a prominent leash of blood vessels grows into the floor of the phlyctenular ulcer and forms a *fascicular ulcer*. The ulcer progresses towards the center of the cornea with advancing gray infiltrate, while cicatrization occurs at the periphery. The ulcer remains superficial and seldom perforates.

The confluence of multiple phlyctens at the limbus causes a *ring ulcer* which may endanger the cornea. A sectorial superficial dendritic phlyctenular pannus is not infrequent and usually causes intense photophobia and blepharospasm.

The treatment of phlyctenular keratitis is same as that of phlyctenular conjunctivitis. The corneal involvement warrants the use of a cycloplegic agent.

Vernal Keratitis

Vernal keratoconjunctivitis can involve the cornea and produce several types of lesions such as superficial punctate keratitis, punctate epithelial erosions in superior and central cornea, noninfec-

tious oval or shield ulcers with underlying stromal opacification.

The corneal lesions respond to the usual treatment of vernal keratoconjunctivitis.

Neurotrophic Keratopathy (Neurotrophic Corneal Ulcer)

Etiology Neurotrophic keratopathy results from a damage to the trigeminal nerve which supplies the cornea. The sensory nerve helps to maintain the corneal epithelium healthy. The loss of neural reflex leads to hydration and exfoliation of the epithelial cells. Neurotrophic keratopathy can develop despite the normal tear secretion and blink reflex. The common causes of the nerve damage are herpes simplex viral infection, herpes zoster ophthalmicus, leprosy and injection of alcohol in the gasserian ganglion for the treatment of trigeminal neuralgia.

Clinical features The patient remains symptom-free. There is absence of pain and lacrimation in spite of the presence of ciliary injection and multiple corneal erosions. The cornea appears dull and exfoliated. There occurs a complete loss of corneal sensation. A refractory corneal ulcer develops in unattended cases.

Treatment The management of neurotrophic ulcer includes frequent instillations of artificial tears, antibiotic and atropine ointments and protection of the eye either by pad and bandage or bandage contact lens. Tarsorrhaphy is a good alternative.

Keratitis Lagophthalmos (Exposure Keratitis)

Etiology Nonclosure or incomplete closure of the palpebral aperture by lids, when eyes are shut, results in exposure keratitis. Bell's palsy, marked proptosis and ectropion are common causes of lagophthalmos.

Clinical features Exposure keratitis is usually seen in the lower part of the cornea. It may range from mild desiccation to suppuration of the cornea which may subsequently perforate.

Treatment The condition can be managed by frequent instillations of tear substitutes in day time and application of eye ointment at night. If corneal ulcer develops, routine treatment of ulcer should be administered. The eye can be protected by the use of bandage contact lens or tarsorrhaphy.

Rosacea Keratitis

Etiology A chronic recalcitrant keratitis is often found associated with acne rosacea.

Clinical features Rosacea is a chronic skin disease characterized by butterfly-like erythema of cheeks and nose associated with telangiectasia, hypertrophy of sebaceous glands, corneal infiltrates and vascularization. Rosacea keratitis is usually associated with acneform lesions of the face and seborrheic blepharitis.

The patient complains of irritation and mild redness of the eyes. The conjunctival vessels in the interpalpebral region are dilated and small gray nodules appear near the limbus which may ulcerate and invade the cornea. Gradually, the cornea gets vascularized. The ulcers are indolent and frequently recur causing irregular corneal facets. Other corneal lesions include map-dot subepithelial opacities, punctate epithelial keratopathy involving the lower two-thirds, recurrent epithelial erosions and thinning of the cornea.

Treatment The treatment of rosacea keratitis is unsatisfactory. The keratitis should be treated on the lines of phlyctenular lesions. Topical corticosteroids and systemic tetracycline (250 mg) four times a day for one month and then once daily for six months or doxycyclin (100 mg) twice daily for 3 weeks may cure both ocular and skin lesions. Superficial X-ray therapy is claimed to be beneficial.

Deep Keratitis

Interstitial keratitis is the most frequent type of deep keratitis. Other forms of deep keratitis include disciform keratitis, keratitis profunda and sclerosing keratitis.

Interstitial Keratitis

Interstitial keratitis is a parenchymatous inflammation of the cornea, more often of allergic origin, wherein the corneal stroma is secondarily involved due to a primary anterior uveitis. The disease frequently affects children suffering from congenital syphilis. It may also be seen in acquired syphilis, tuberculosis, sarcoidosis, leprosy, trachoma, Lyme disease, mumps, brucellosis, trypanosomiasis, onchocerciasis and malaria.

Syphilitic Interstitial Keratitis

Etiology Syphilitic interstitial keratitis occurs in two forms: congenital and acquired. The former is more frequent.

Congenital infection with *Treponema pallidum* can occur via transplacental route. The disease is usually bilateral (80%) and affects the children between the ages of 5 and 15 years.

Pathogenesis In interstitial keratitis, anterior uvea is almost always affected as evidenced by the presence of keratic precipitates. The basic lesion is uveitis, and keratitis is the result of immune-mediated reaction. *Treponema pallidum* is not seen in the cornea even during the acute phase.

In the initial phase, cellular infiltration appears in the deeper layers of the cornea just anterior to Descemet's membrane. The characteristic cell is lymphocyte. There is always some edema of the corneal epithelium and endothelium causing thickening and cloudiness of the tissue. The corneal lamellae get separated and undergo necrosis. Meanwhile, blood vessels from the limbus grow in a brush-like manner and invade the deeper layers of the cornea. The dense infiltration produces folds in Descemet's membrane and, sometimes, result in destruction of this layer and underlying endothelium. Bowman's membrane may also undergo destruction. The debris is removed by macrophages and healing occurs by proliferation of corneal fibroblasts which convert the necrosed area into a vascularized scar. The regression occurs slowly, the corneal edema disappears and the vessels start obliterating. However, ghost vessels remain throughout the life as fine lines despite the resolution of the disease.

Clinical features Syphilitic or leutic interstitial keratitis often follows an injury or an operation on the eye. The clinical course of the disease may be divided into three stages: progressive, florid and regressive.

Progressive stage: An indistinct cellular infiltrate or stromal opacities, mild endothelial edema and KPs constitute initial notable signs of an active disease. Ciliary injection develops later. Irritation, watering, photophobia and pain are usual accompanying symptoms. The disease often begins in the periphery and involves the upper part of the cornea initially. The discrete infiltrate in the stroma enlarges and spreads towards the center of the cornea and eventually renders the entire cornea a dull and cloudy appearance. The cornea assumes a typical ground glass appearance in 2-4 weeks.

Florid stage: A dense infiltration and vascularization of the corneal stroma develop in this stage. The vascular growth begins at the periphery and remains sectorial. The deep radial vessels are arranged in a brush-like fashion and look dull reddish-pink (Fig. 12.25) owing to the overlying hazy cornea (*Salmon patch of Hutchinson*). The superficial conjunctival vessels are congested but never extend far over the cornea. However, there occurs an epaulette-like heaping of the conjunctiva at the limbus.

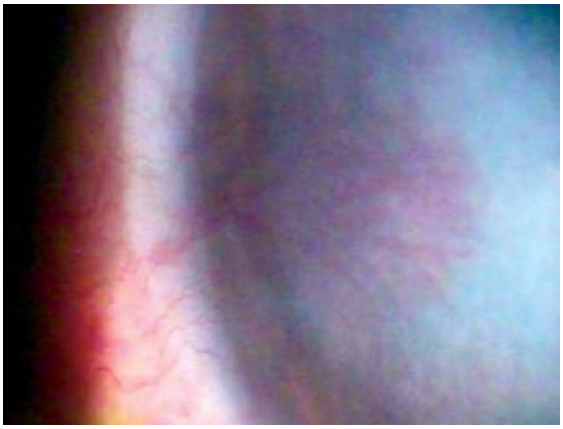


Fig. 12.25: Deep vascularization in syphlitic interstitial keratitis

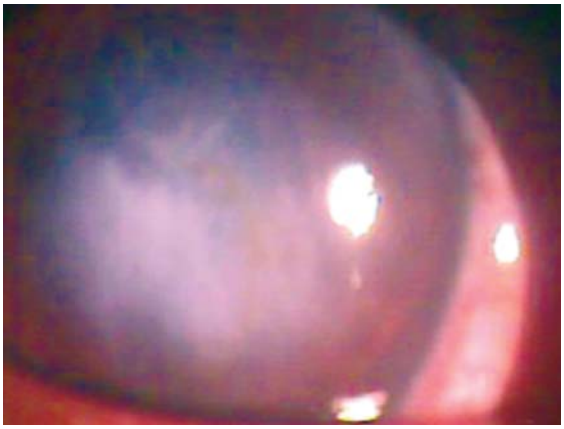


Fig. 12.26: Tuberculous interstitial keratitis

Regressive stage: Once the entire cornea has vascularized, the regressive stage begins. During this stage the symptoms are minimum except marked impairment of vision. Gradually, the cornea starts clearing from the periphery towards the center. The stromal vessels begin to recede and vision improves slowly. The cornea shows a few deep opacities and empty or ghost vessels. If the cornea does not clear up within 18 months, the visual prognosis is usually poor.

Children with interstitial keratitis may have nonocular signs of syphilis. Stigmata of congenital syphilis include frontal prominence, depressed bridge of the nose, Hutchinson's teeth, vestibular deafness and rhagades at the angle of mouth.

The diagnosis of syphilis can be confirmed serologically with rapid plasmin reagent test and fluorescent treponemal antibody absorption (FTA-ABS) test.

Tuberculous Interstitial Keratitis

Tuberculous interstitial keratitis (Fig. 12.26) is usually unilateral and involves the lower half or a sector of the cornea. The clinical features are almost same as found in the syphilitic interstitial keratitis. Primary complex may be present in some cases.

Treatment Both local and systemic treatment should be started in the initial stage of the disease to obtain good results. As interstitial keratitis is a type IV hypersensitivity response to microorganism, prednisolone 1% eye drops should be instilled every 2 hourly. Topically cycloplegics (atropine 1%) must be applied 2 to 3 times a day to give rest to the anterior uvea.

Refractory cases often need more energetic treatment with subconjunctival or sub-Tenon injections of corticosteroids. Hot compresses are soothing and dark glasses relieve photophobia.

Systemic corticosteroids should always be combined with antisyphilitic or antitubercular therapy. The treatment should not be terminated abruptly, otherwise recurrence may occur. With prompt and adequate therapy, the cornea may clear up with recovery of useful vision. In severe cases, where dense corneal opacities are formed, good results can be obtained with penetrating keratoplasty.

Keratitis Profunda

Etiology Keratitis profunda is a deep type of keratitis having an obscure etiology. It affects adults and is often unilateral and may be associated with iridocyclitis.

Clinical features Pain, photophobia, lacrimation and diminution of vision are usual symptoms. The clinical signs include ciliary congestion,

corneal epithelial edema, deep stromal opacity, folds in Descemet's membrane and mild deep vascularization.

Treatment Topical cycloplegics and corticosteroids improve the condition.

Disciform Keratitis

Disciform keratitis is characterized by the appearance of a disk-shaped dense grayish infiltration in the deeper layers of the central cornea.

Etiology Disciform keratitis is often associated with viral infection. The condition is usually unilateral and may be caused by herpes simplex virus, adenoviruses and trauma. Disciform keratitis is considered as an allergic response of the stroma to the virus.

Clinical features The signs and symptoms of disciform keratitis are akin to that seen in herpes simplex keratitis. The vision is considerably impaired due to central location of the opacity. The cornea becomes anesthetic.

Treatment Topical acyclovir 3% along with the use of topical corticosteroids may be beneficial in selected cases.

Sclerosing Keratitis

Sclerosing keratitis is a complication of scleritis. It may accompany herpes zoster scleritis or rheumatoid arthritis.

Clinical features A tongue-shaped opacity develops at the margin of cornea adjacent to a patch of scleritis. The rounded apex of the tongue is directed towards the center of cornea. The opacity is composed of grayish lymphocytic infiltrates in the stroma resembling the sclera, hence the term sclerosing keratitis. Developmentally, the stroma of the cornea is a differentiated part of the sclera, therefore, its preferential involvement in the affection of the sclera occurs. Sclerosing keratitis has a little or no vascularization and it never ulcerates. The opacity clears from the center towards the periphery of the cornea but some cloudiness persists in the zone of deep infiltration.

Treatment Timely treatment of scleritis helps in resolution of sclerosing keratitis.

DEGENERATIVE CONDITIONS OF THE CORNEA

Cornea is a common site for infiltrations associated with inborn errors of lipid and mucopolysaccharide metabolisms and also for the primary and secondary degenerations.

Arcus Senilis (Anterior Embryotoxon)

Arcus senilis is a lipid infiltration of the peripheral corneal stroma often seen in old age (Fig. 12.27). It is inherited as an autosomal dominant trait. Arteriosclerosis, hyperlipidemia, hypercholestrolemia and coronary artery disease are implicated in the etiology of arcus senilis.

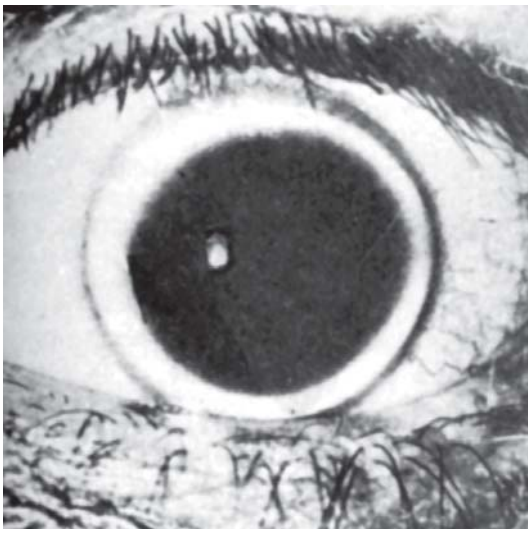


Fig. 12.27: Arcus senilis

The selective infiltration of lipid in the periphery of the cornea leaves a clear area between the limbus and the arcus which is known as *lucid interval*. The arcus usually commences as a crescent preferentially at the upper and the lower margin of the cornea. Later the extremities of the crescent join to form a circle. It is mostly limited to the periphery of the cornea but sometimes it may cover 2 to 3 mm of the periphery; in spite of the spread it does not affect the vision.

Arcus Juvenilis

The appearance of an arcus in young persons is known as *arcus juvenilis*. It may be associated with megalocornea, keratoconus, vernal keratoconjunctivitis and familial lipidemia.

Hassall-Henle Bodies

Localized nodular thickenings in the periphery of Descemet's membrane are known as *Hassall-Henle bodies*. They appear as small dark areas within the normal endothelial mosaic. They are caused by over production of hyaline by the endothelial cells and represent ageing process.

Spheroidal Degeneration or Climatic Droplet Keratopathy

Spheroidal degeneration or climatic droplet keratopathy is characterized by subepithelial accumulation of opalescent droplets that coalesce to form bands or nodules with elevated corneal epithelium. The degeneration is mainly seen in the interpalpebral portion of the cornea. Probably it is caused by microtrauma from wind and sand or solar radiation. Excimer laser keratectomy or lamellar keratoplasy helps improving the vision.

Salzmann's Nodular Degeneration

Etiology A nodular degeneration of the cornea may occur following recurrent attacks of phlyctenular keratoconjunctivitis. It may also occur as a late sequel to trachoma, vernal keratoconjunctivitis and measles.

Clinical features The corneal surface, particularly near the limbus, may show a chain of bluish-white elevated avascular nodules. The degeneration may cause irritation and impairment of vision.

Treatment Lamellar keratoplasty improves the vision.

Band-shaped Keratopathy

Etiology A transverse band-shaped opacity of the cornea (Fig. 12.28) with calcium deposits may be found in juvenile rheumatoid arthritis (Still's disease), chronic anterior uveitis, sarcoidosis, leprosy, absolute glaucoma, phthisical eye, hyperparathyroidism and vitamin D intoxication.

Clinical features Irritation, lacrimation and diminution of vision are the presenting symptoms. Band-shaped keratopathy is limited to the interpalpebral area and there exists a clear corneal strip between the band and the limbus. Bowman's membrane and the anterior stroma are destroyed. Subsequently fibrosis and deposition of calcareous material take place.

Treatment The treatment of band-shaped keratopathy consists of removal of the corneal epithelium

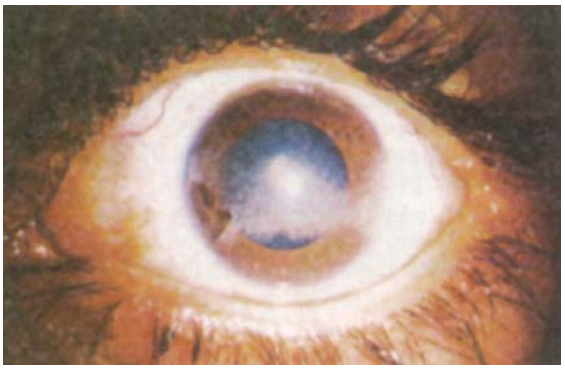


Fig. 12.28: Band-shaped keratopathy

followed by application of 0.01 M solution of ethylene diamine tetraacetic acid (EDTA) or neutral ammonium tartarate. Excimer laser keratectomy (phototherapeutic keratectomy) can be helpful in improving the vision. Some cases need penetrating keratoplasty.

Hereditary Corneal Dystrophies

Corneal dystrophies are bilateral symmetrical inherited conditions which involve the central part of the cornea. Dystrophies begin early in life and tend to be slowly progressive.

Classification Corneal dystrophies can be classified into 2 broad groups:

1. **Anterior corneal dystrophies**
   - **A.** ***Epithelial corneal dystrophies***
     - i. Cogan's microcystic dystrophy
     - ii. Meesmann's dystrophy
     - iii. Reis-Buckler dystrophy
     - iv. Thiel-Behnke dystrophy.
   - **B.** ***Stromal corneal dystrophies***
     - i. Granular
     - ii. Macular
     - iii. Lattice
   - **C.** ***Endothelial corneal dystrophies***
     - i. Endothelial corneal dystrophy (Fuchs)
     - ii. Posterior polymorphous dystrophy
2. **Ectatic corneal dystrophies**
   - i. Keratoconus
   - ii. Keratoglobus
   - iii. Pellucid marginal degeneration

The corneal epithelium and Bowman's layer are affected in epithelial corneal dystrophy; the stroma and Descemet's membrane are involved in stromal corneal dystrophy and the endothelium in endothelial corneal dystrophy. Ectatic corneal dystrophies cause weakness of the entire cornea leading to change in its curvature.

Epithelial Corneal Dystrophies

Cogan's Microcystic Dystrophy (Corneal Epithelial Basement Membrane Dystrophy)

Cogan's microcystic dystrophy is the most common of all corneal dystrophies. It is characterized by bilateral, cystic, dot-like or linear fingerprint-like lesions in the corneal epithelium. The pattern of lesions and their distribution may vary with time. The condition is asymptomatic. However, after the age of 30 years, some patients may develop recurrent corneal erosions precipitated by trivial trauma. The main defect lies in the basement membrane of the epithelium.

Meesmann (Juvenile Epithelial) Dystrophy

Meesmann dystrophy is a rare autosomal dominant epithelial dystrophy which manifests early in life. Mild irritation and decrease in visual acuity are the presenting symptoms. Small bubble-like blebs are seen in the interpalpebral area. Occasionally, epithelial lesions take the shape of whorls or wedges. Corneal sensation is usually reduced.

Reis-Buckler Dystrophy

Reis-Buckler corneal dystrophy is an autosomal dominant dystrophy which appears in the first few years of life. It is characterized by a superficial geographic or homogenous gray-white reticular or fish-net pattern opacification of the central cornea associated with impairment of vision. The recurrent epithelial erosions of the cornea cause pain and watering.

Theil-Behnke Dystrophy

Theil-Behnke dystrophy is a Bowman's membrane dystrophy presenting as a honeycomb

opacification of the superficial central cornea. It demonstrates curly fibers on electron microscopy.

Treatment of epithelial dystrophies includes application of 5% sodium chloride drops 5 times a day and sodium chloride 6% ointment at bed time, epithelial debridement, soft contact lens and phototherapeutic keratectomy (PTK).

Stromal Corneal Dystrophies

There are three main types of stromal corneal dystrophies.

Granular Corneal Dystrophy

Granular (*Groenouw type I*) is the most common stromal dystrophy. It is inherited as an autosomal dominant trait and usually presents during the first decade of life. Multiple discrete crushed bread crumb-like white granular opacities develop in the axial region of the anterior corneal stroma (Fig. 12.29), the peripheral cornea is seldom involved. The granular material is eosinophilic hyaline in nature and stains bright red with Masson trichrome stain.

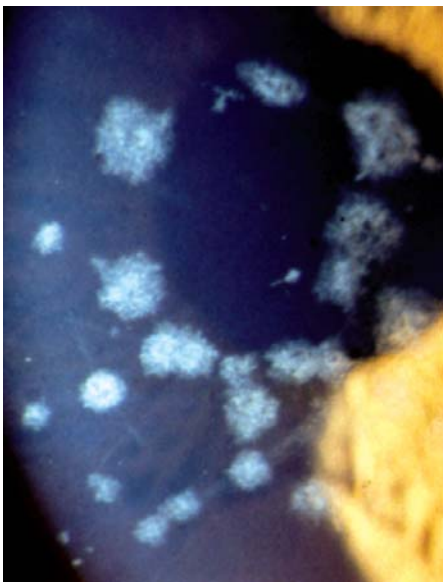


Fig. 12.29: Granular corneal dystrophy (*Courtesy*: Mr S Kanagami,Tokyo)

Vision is good despite the presence of some glare. The opacities coalesce into various irregular forms to jeopardize the vision in the fourth decade. Contact lens, phototherapeutic keratectomy and penetrating keratoplasty can restore the vision.

Macular Corneal Dystrophy

Macular (*Groenouw type II*) dystrophy is inherited in an autosomal recessive manner and involves central as well as peripheral parts of the corneal stroma. Numerous grayish, poorly defined opacities commence in the axial cornea and then spread to the corneal periphery (Fig. 12.30). The nature of deposit in macular corneal dystrophy is glycosaminoglycan (GAG) which stains with colloidal iron and Alcian blue. Macular dystrophy affects the vision at an early age. Corneal sensation is usually impaired and irritation and watering are common symptoms due to recurrent corneal erosions. Lamellar keratoplasty is indicated for the management of macular corneal dystrophy.

Lattice Corneal Dystrophy

Lattice (*Biber-Haab-Dimmer*) dystrophy is inherited as an autosomal dominant trait and manifests during the latter part of the first decade as recurrent corneal erosions. It is characterized by spider-like lines in the corneal stroma. The branching

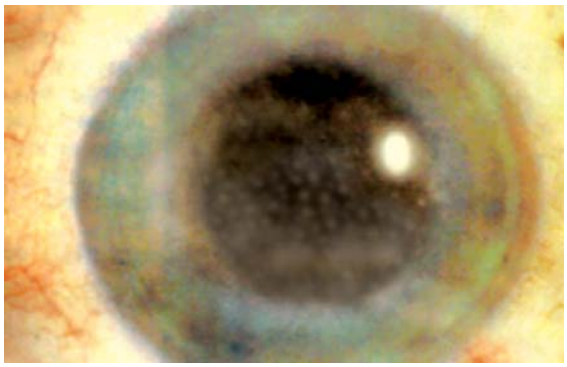


Fig. 12.30: Macular corneal dystrophy

filaments are arranged in radial pattern interlacing at different levels causing impairment of vision. Amyloid deposit occurs in the corneal stroma. Amyloid stains rose to orange red with Congo-red dye. The corneal sensation is impaired. Corneal grafting restores the vision.

Endothelial Corneal Dystrophies

Fuchs Endothelial Corneal Dystrophy

Fuchs endothelial dystrophy (FED) is an autosomal dominant condition occurring after 50 years of age with a female predominance. The severity of the disease varies from asymptomatic corneal guttata to markedly decompensated cornea. Corneal guttata (Fig. 12.31) first appears centrally and then spreads towards the periphery. Descemet's membrane folds develop secondary to stromal edema. Decompensation of the endothelium causes epithelial microcystic edema and epithelial bullae. Rupture of bullae causes pain.

Specular microscopy is helpful in the diagnosis of Fuchs dystrophy. When the endothelial cell count is less than 1000/mm^2 or the corneal thickness is greater than 650 µm, extra precautions during the intraocular surgery should be taken to protect the endothelium from surgical trauma.

Use of sodium chloride drops (5%) and ointment (6%) and oral carbonic anhydrase inhibitor may reduce the edema. Lubricating drops and soft contact lenses relieve pain caused by rupture of bullae. Keratoplasty can restore the vision.

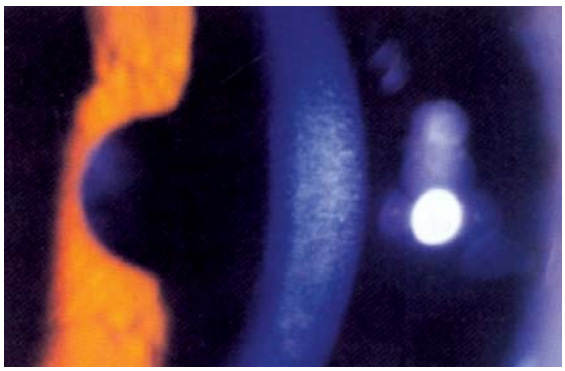


Fig. 12.31: Fuchs endothelial dystrophy

Posterior Polymorphous Dystrophy

Posterior polymorphous dystrophy (PPMD) is an uncommon dystrophy that occurs early in life. The characteristic microscopic feature of the dystrophy is the presence of multilayered endothelial cells that behave like fibroblasts. The posterior surface of cornea shows vesicles and gray broad bands. Stromal micropuncture or penetrating keratoplasty can improve the condition.

Ectatic Corneal Dystrophies

Keratoconus

Keratoconus is a common curvature disorder of the cornea in which the central or paracentral cornea undergoes a progressive thinning or bulging taking the shape of a cone (Fig. 12.32).

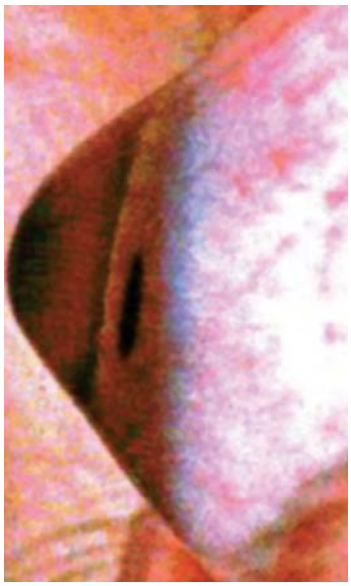


Fig. 12.32: Keratoconus

Etiology The etiology of keratoconus is unknown but it seems to be multifactorial. It is a familial disorder showing female predominance. A locus of keratoconus has been identified on chromosome 21. Down's syndrome has a close association with keratoconus.

Clinical features Keratoconus is a bilateral and asymmetrical curvature anomaly of the cornea which often progesses slowly and manifests at puberty causing marked visual impairment. It presents a scissor red reflex on retinoscopy (*Rizzutti's sign*) which is a very early sign of keratoconus. The visual loss in keratoconus occurs due to irregular astigmatism and corneal scarring.

There occurs a conical protrusion of the cornea, the apex of the cone being slightly below the center of the cornea. A conical reflection on the nasal cornea is seen when light is shown from the temporal side. The alteration in the curvature of the cornea produces distortion of the corneal reflex as seen with the Placido's disk or on corneal topography (Fig. 12.33). When the patient looks down, an indentation in the lower lid by the cone of the cornea may be noticed (*Munson's sign*). Slit-lamp biomicroscopy reveals thinning and opacities at the apex of cornea, increased visibility of the corneal nerves and a brownish ring at the base of cone, probably due to deposition of hemosiderin in the corneal epithelium (*Fleischer ring*). An absence of Bowman's membrane and presence of stress lines (*Vogt striae*) in the stroma may also be seen.

A tear in Descemet's membrane causes acute hydrops resulting in a sudden impairment of vision (which may be regained spontaneously) associated with moderate ocular pain and corneal edema. Rarely perforation may occur.

Treatment Initially all patients with keratoconus should be prescribed glasses or rigid gas permeable contact lenses to correct the refractive error. Hydrops should be treated conservatively by frequent instillations of hyperosmotic agents and cycloplegics, and discarding contact lens wear. Penetrating keratoplasty provides good and lasting visual results.

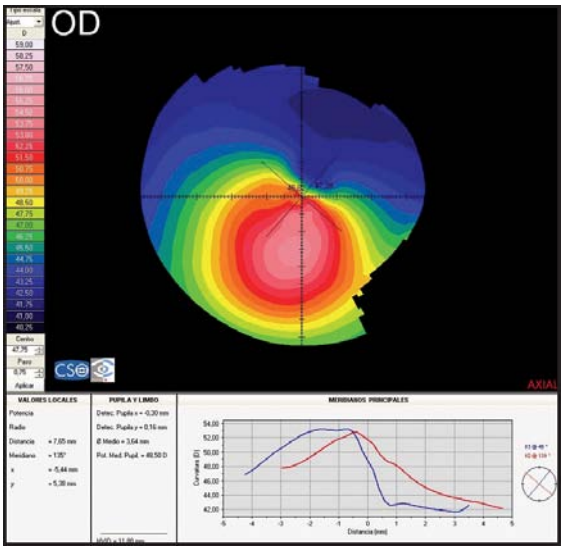


Fig. 12.33: Corneal topography of keratoconus

Posterior Keratoconus

Posterior keratoconus is a unilateral, congenital, nonprogressive condition characterized by a localized or generalized defect of the posterior surface of the cornea with concavity towards the anterior chamber.

Keratoglobus

Keratoglobus is a congenital curvature anomaly of the entire cornea in which the cornea assumes a hemispherical shape. It represents a defect in collagen synthesis and is inhertited as an autosomal recessive trait.

Keratoglobus is a bilaterally symmetrical nonprogressive condition. In spite of the thinning, the cornea remains clear. The presence of normal intraocular pressure, open angle of the anterior chamber and absence of cupping of the optic disk differentiates keratoglobus from buphthalmos. A

central stromal haze (fragmentation of Bowman's membrane) is often present, but apical scar, stress lines and Fleischer's ring are absent. Blue sclera and hyperextensibility of hand and ankle joint may be associated with keratoglobus.

Pellucid Marginal Degeneration

Pellucid marginal degeneration is a non-hereditary bilateral inferior peripheral corneal thinning seen in young patients. The etiology of the condition is unknown.

Protrusion of the cornea occurs above the band of thinning causing a decrease in vision due to high astigmatism. Vascularization of the cornea does not occur but posterior stromal scarring has been noted. Sometimes, acute hydrops may develop.

The patient usually needs correction for astigmatism by contact lenses. Some patients may require lamellar tectonic graft.

CONGENITAL ANOMALIES OF THE CORNEA

The congenital anomalies of cornea may result either from the disturbances in the process of formation or differentiation of the individual layer. Anomalies of transparency and curvature are not uncommon. Microcornea, cornea plana, megalocornea, Peter's anomaly and corneal dermoid are some of the congenital anomalies of the cornea.

Microcornea

Microcornea (Fig. 12.34) may be an isolated anomaly or it may accompany other anterior segment defects. The size of the cornea is less than 10 mm. The cornea is usually clear and flat and prone for the development of angle-closure glaucoma.

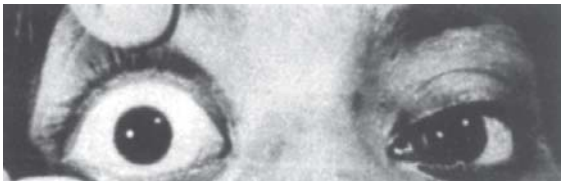


Fig. 12.34: Unilateral microcornea

Cornea Plana

Cornea plana is a rare congenital defect wherein the corneal curvature is markedly reduced. It may be associated with scleralization of the cornea or infantile glaucoma.

Megalocornea

Megalocornea is a developmentally enlarged cornea measuring more than 13 mm in horizontal diameter. The cornea is transparent and histologically normal. This bilateral anomaly is transmitted in a sex-linked recessive manner, mostly affecting males (90%).

Peter's Anomaly

Peter's anomaly is characterized by a triad of abnomalities: central or paracentral posterior corneal defect with overlying corneal opacities, iridocorneal adhesion to the edge of the defect and corneo-lenticular contact or cataract.

Corneal Dermoid

Corneal dermoid is a unilateral or bilateral tumor present at birth and commonly involves the limbus. It is composed of choristomatous tissue and often enlarges at puberty. It is a small, discrete, slightly elevated, firm, white-yellow translucent mass usually straddling the limbus and occupying a part of the cornea, and rarely may replace the entire cornea. It may constitute a part of Goldenhar syndrome (*oculo-auriculo-vertebral syndrome*).

SYMPTOMATIC CONDITIONS OF THE CORNEA

Corneal Edema

The corneal endothelium maintains the transparency of cornea by acting as a barrier membrane and by providing a metabolic pump. Whenever the selective permeability of the corneal endothelium and epithelium is impaired and the endothelium fails to pump-out water, hydration of corneal stroma and epithelium occurs thereby affecting the corneal transparency. Besides keratitis, uveitis, trauma (surgical or otherwise), endothelial dystrophy and raised intraocular pressure cause corneal edema. Wearing of contact lenses for extended period may also produce corneal edema.

Clinical features Ocular discomfort, watering and impairment of vision are common complaints of the patient. The cornea appears hazy (Fig. 12.35). Decompensated cornea presents deep irregular stromal opacities associated with epithelial bullae.

Treatment The corneal edema can be managed by treating the primary cause. Sodium chloride 5% drops 5 times daily and sodium chloride 6% ointment at night give relief. Bandage contact lens can minimize the discomfort. Long-standing decompensated cornea needs penetrating keratoplasty.

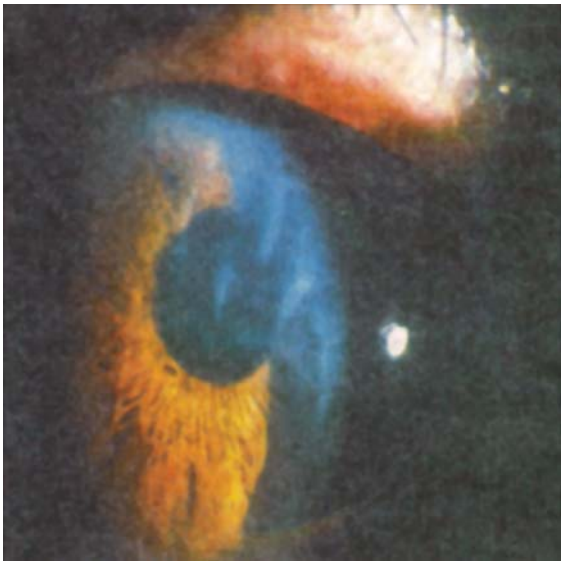


Fig. 12.35: Corneal edema

Corneal Opacities

Etiology Congenital corneal opacities are uncommon. However, striate opacities are frequent following intraocular surgery, especially after cataract extraction. They are due to the wrinkling of Descemet's membrane and adjoining stroma, and appear as fine gray lines radiating from the wound and running across the cornea. They disappear with the healing of the wound.

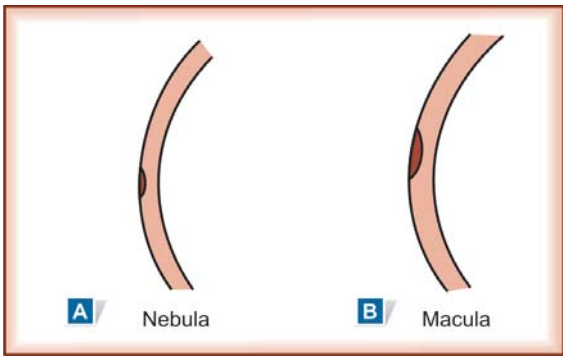
Permanent corneal opacities are due to corneal ulcer, deep keratitis, dystrophy or degeneration. The corneal tissue is destroyed and replaced by disorderly arranged fibrous lamellae covered with thick irregular epithelium.

Clinical features Visual disturbances and cosmetic disfigurement are frequent symptoms. The visual impaiment caused by a corneal opacity may vary depending on its site and density. The opacity situated in the periphery of the cornea does not affect the vision while centrally located one causes significant impairment. The opacity may or may not be vascularized.

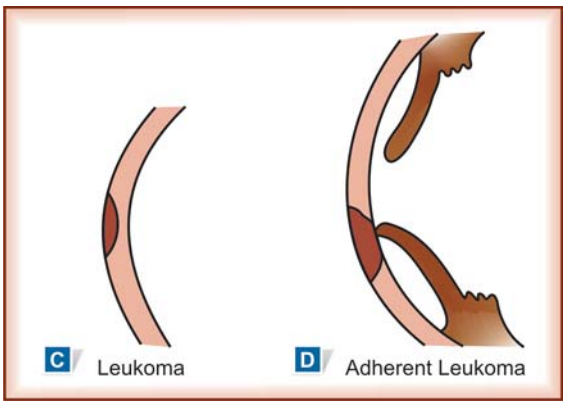
Depending on the density, corneal opacities are graded as nebula, macula and leukoma.

Nebular corneal opacity: When the corneal scar is thin it is known as *nebula* (Fig. 12.36A). It is usually caused by destruction of Bowman's membrane and superficial stroma. The opacity may be so faint that it can be missed on routine examination unless cornea is examined on a slit-lamp. Nebula may be localized or diffuse. Presence of nebula in the pupillary area causes blurring of vision due to irregular astigmatism.

Macular corneal opacity: When the corneal scar is moderately thick, it is called *macula* (Fig. 12.36B).



Figs 12.36A and B: Corneal opacity: (A) Nebula, (B) Macula



Figs 12.36C and D: Corneal opacity: (C) Leukoma, (D) Adherent leukoma

It is caused by destruction of less than half the thickness of corneal stroma.

Leukomatous corneal opacity: When the opacity is very dense and white, it is called *leukoma* (Fig. 12.36C). The destruction of more than half the thickness of corneal stroma causes leukomatous opacity. Occasionally the corneal scar is weak and thin and bulges under the normal intraocular pressure, the condition is known as *keratectasia.*

When iris tissue is adherent to the back of a leukoma, it is called as *leukoma adherence* (Fig. 12.36D), a common sequel of a perforated corneal ulcer.

The perforation of a sloughing corneal ulcer results in formation of a pseudocornea over the prolapsed iris. The ectasia of pseudocornea with the incarceration of iris tissue is known as *anterior staphyloma*. It may be partial or total. The anterior chamber is absent in total anterior staphyloma. The intraocular pressure is often raised due to the development of secondary glaucoma. The anterior staphyloma may appear as conical, globular or lobulated (Fig. 12.37). The leukomatous corneal opacity has brown or slaty discolouration representing incarceration of the iris tissue. The condition is not only disfiguring but also painful. The summit of the staphyloma may get ulcerated and the aqueous humor may leak out. Vascularization on the surface of staphyloma is not uncommon.

Treatment There is no satisfactory medical treatment for corneal opacity. When the corneal opacity covers the pupillary area an optical iridectomy can improve the vision, but the ideal procedure is either excimer laser phototherapeutic keratectomy or corneal transplantation.

Temporary cosmetic improvement may be obtained by *tattooing* the corneal opacity with gold chloride or platinum chloride. The epithelium over

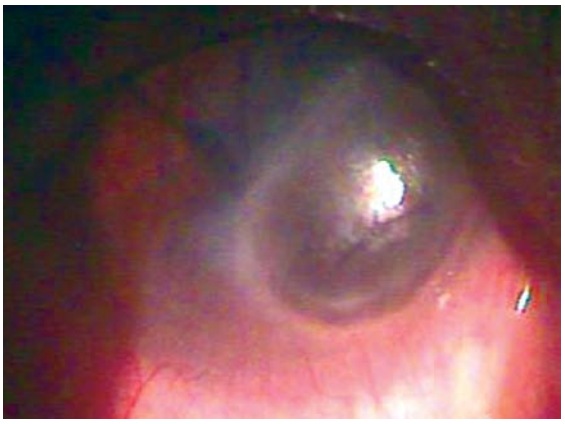


Fig. 12.37: Anterior staphyloma

the opacity is scraped off after anesthetizing the surface. Gold chloride 4% or platinum chloride 2% solution is applied for 2 to 3 minutes to impregnate the scar. A freshly prepared hydrazine hydrate 2% solution is instilled over the cornea to reduce gold chloride to dark brown and platinum chloride to black color. The eye should be bandaged after application of atropine ointment.

A partial anterior staphyloma is managed by reducing the intraocular pressure and performing penetrating keratoplasty. A total staphyloma is dealt with enucleation or staphylectomy.

Vascularization of the Cornea

The cornea is an avascular tissue and presence of blood vessels in the cornea is always pathological. Superficial vascularization (Fig. 12.38) and deep vascularization (Fig. 12.25) appear in inflammatory disorders of the cornea.

The superficial vascularization of cornea is common in trachoma, superficial corneal ulcers, phlyctenular keratoconjunctivitis, rosacea keratitis and contact lens wearers.

The deep vascularization of cornea is seen in interstitial keratitis, deep corneal ulcers, sclerosing keratitis, disciform keratitis and chemical burns.

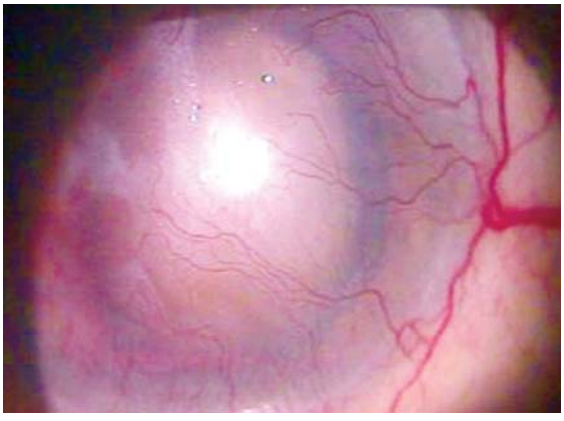


Fig. 12.38: Superficial vascularization of cornea

The vascularization of cornea may be prevented by timely and adequate treatment of predisposing conditions. Application of topical corticosteroids, thiotepa or β-irradiation is effective. Intractable cases are dealt with peritomy or corneal grafting.

Pigmentation of the Cornea

Pigmentation of the cornea may be due to prolonged use of topical drugs, trauma, foreign body, inborn errors of metabolism and degeneration.

Iatrogenic pigmentation of the cornea may occur from prolonged use of silver nitrate. Repeated silver nitrate application leads to brownish discoloration of Descemet's membrane owing to impregnation of the salt (*argyrosis*). Prolonged topical application of epinephrine in the management of glaucoma may result in black cornea.

A retained copper foreign body in the eye may produce a grayish-green or golden-brown discoloration of the peripheral corneal stroma (*chalcosis*). Deposition of copper between Descemet's membrane and corneal endothelium is found in hepatolenticular degeneration (Wilson's disease) where a green or brown ring is seen just inside the limbus (*Kayser-Fleischer ring*). The intensity of pigmentation can be reduced by administration of penicillamine.

Blood staining of the cornea can follow massive hyphema either from a contusion injury or an intraocular surgery. A rise of intraocular pressure often promotes blood staining of the cornea. The deeper layers of cornea are stained with blood pigment (hemosiderin) and may develop brown or greenish discoloration simulating dislocation of the lens in the anterior chamber. The cornea very slowly clears from periphery towards the center.

A brown horizontal line (*Hudson-Stahli line*) in the inferior third of the cornea may be seen on slit-lamp in elderly persons. Similarly, a vertical spindle-shaped brown uveal pigments deposition

on the corneal endothelium (*Krukenberg's spindle*) may be found in a small percentage of myopic eyes. The spindle may be associated with pigment dispersion glaucoma.

Fleischer's ring, represents deposition of hemosiderin in the corneal epithelium, is often found in keratoconus.

Stocker's line is a golden brown line in the corneal epithelium located at the leading edge of a pterygium representing deposition of iron.

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CHAPTER

13 Diseases of the Sclera

ANATOMY

The word sclera is derived from a Greek word meaning hard. It forms the posterior five-sixths part of the fibrous outer protective tunic of the eyeball. Its outer surface is in contact with Tenon's capsule and the bulbar conjunctiva. The sclera is covered by a thin layer of loose tissue called *episclera*. It is separated from the choroid by the suprachoroidal space. The extraocular muscles are inserted in the sclera.

The thickness of the sclera varies from place to place. The thickest part is at the posterior pole and the thinnest underneath the insertion of rectus muscles. The sclera thins out at the equator.

At the entrance of the optic nerve, the sclera is modified into a sieve-like membrane, the *lamina cribrosa*, which allows the passage of fasciculi of the nerve. The sclera is pierced by two long and ten to twelve short posterior ciliary arteries around the optic nerve. Slightly posterior to the equator, four vortex veins (*venae vorticosae*) exit through the sclera. The anterior ciliary arteries and veins penetrate the sclera nearly 3 to 4 mm away from the limbus.

Histologically, the sclera consists of three layers from without inwards (Fig. 13.1), episcleral tissue, sclera proper and lamina fusca.

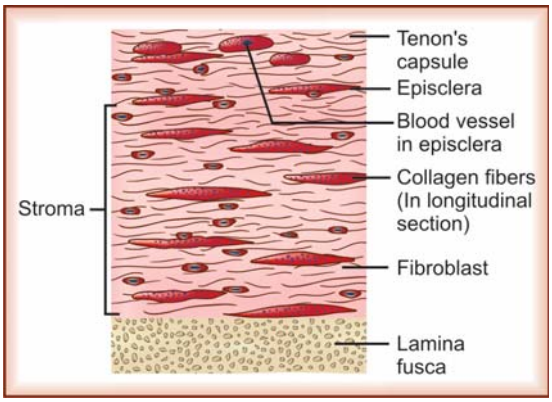

Fig. 13.1: Diagram showing layers of sclera

The *episcleral tissue* comprises fine loose elastic tissue fibers and contains a large number of small vessels.

The *sclera proper* is formed by dense bands of parallel and interlacing collagen fibers. The collagen fiber bundles are arranged in concentric circles at the limbus and around the entrance of the optic nerve, elsewhere the arrangement is quite complicated.

The *lamina fusca* has a brown color owing to the presence of a large number of branched chromatophores.

The sclera is almost avascular and its histological structure resembles that of the cornea. However, sclera is opaque due to the hydration and irregular arrangement of its lamellae. The nerve supply of sclera comes through the ciliary nerves.

INFLAMMATION OF THE SCLERA

An inflammation of the sclera is often endogenous in origin and manifests in two distinct forms: episcleritis and scleritis.

Episcleritis

A self-limiting, transient inflammatory involvement of the superficial layers of the sclera is known as *episcleritis*. The condition may be unilateral (more than 60%) or bilateral, predominantly affecting the young women.

Etiology The precise cause is not known but it is considered to be a hypersensitivity reaction to an endogenous tubercular or streptococcal toxin. Episcleritis may be associated with rheumatoid arthritis, polyarteritis nodosa, spondyloarthropathies and gout. There occurs a localized lymphocytic infiltration of episcleral tissue associated with edema and congestion of the conjunctiva and Tenon's capsule.

Episcleritis manifests in two forms—nodular and diffuse.

Clinical features Redness, ocular discomfort or occasional pain, photophobia and lacrimation are the usual symptoms.

Nodular Episcleritis

There occurs a pink or purple circumscribed flat nodule situated 2 to 3 mm away from the limbus, often on the temporal side (Fig. 13.2). It is hard, tender, immobile and the overlying conjunctiva moves freely over it. The episcleral vascular congestion imparts a bright red or salmon pink color to it. The nodule seldom undergoes suppuration or ulceration.

Diffuse Episcleritis

The inflammatory reaction is confined to one or two quadrants of the eye in diffuse episcleritis.

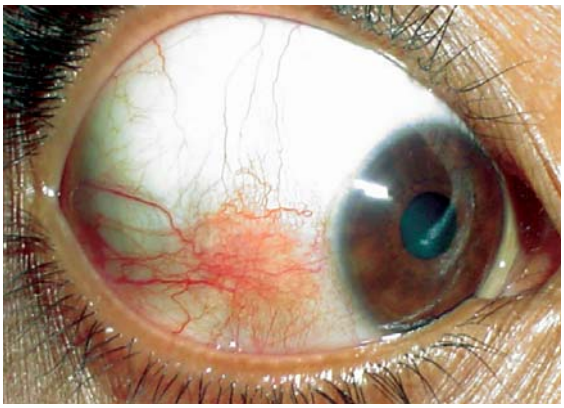

Fig.13.2: Nodular episcleritis

The involved area looks markedly congested. The condition is benign and the course is usually self-limiting. However, recurrences are frequent.

Occasionally, a fleeting type of episcleritis, *episcleritis periodica fugax*, may be seen. The remission of nodule sometimes leaves a translucent sclera. Episcleritis seldom causes scleritis or iritis.

Treatment Topical and oral NSAIDs is the treatment of choice. Severe or recurring disease needs a short course of topical corticosteroids.

Scleritis

Scleritis is a chronic inflammation of the sclera proper often associated with systemic diseases.

Etiology Scleritis is caused by an immune-mediated vasculitis that may lead to destruction of the sclera. It occurs in older age group and affects females more than males. Herpes zoster is the most important local cause of scleritis. Scleritis is frequently associated with connective tissue or autoimmune diseases, especially rheumatoid arthritis (1:200 patients). Sarcoidosis, Behçet's disease, ankylosing spondylitis, gout, tuberculosis and syphilis are also implicated in the etiology of scleritis. Physical, chemical or mechanical injuries are some of the risk factors.

Like episcleritis, scleritis is also considered as a focal hypersensitivity reaction to endogenous toxins as evident by a massive lymphocytic infiltration of the deeper layers of sclera. The initial inflammatory process is caused by immune complex-related vascular damage (type III hypersensitivity reaction) followed by a granulomatous response (type IV hypersensitivity reaction). The scleral stroma becomes necrotic and is replaced by thin fibrous tissue. Some areas of avascularity suggest occlusive vascular phenomenon. The damaged or weakened area may become ectatic, forming a staphyloma.

Classification Scleritis can be classified on the basis of anatomical location and type of scleral inflammation:

A. Anterior scleritis
   1. Nonnecrotizing scleritis
      a. Nodular
      b. Diffuse
   2. Necrotizing scleritis
      a. With inflammation
      b. Without inflammation (Scleromalacia perforans)

B. Posterior scleritis.

Anterior Nonnecrotizing Scleritis

Nonnecrotizing nodular scleritis (Fig. 13.3) is characterized by the presence of one or more hard, purplish, elevated scleral nodules near the limbus associated with marked inflammatory reaction. Sometimes, the nodules may encircle the cornea in an annular fashion, *annular scleritis*, resulting in a severe damage to the anterior segment of the eye.

Nonnecrotizing diffuse scleritis (Figs 13.4A and B) is a more widespread inflammatory reaction involving a sector of the sclera or complete anterior sclera. It is a painful condition with marked reactive edema and loss of vascular pattern of the sclera.

Clinical features The symptoms of scleritis are more marked than episcleritis. They include pain, redness, photophobia, lacrimation and diminution of vision.

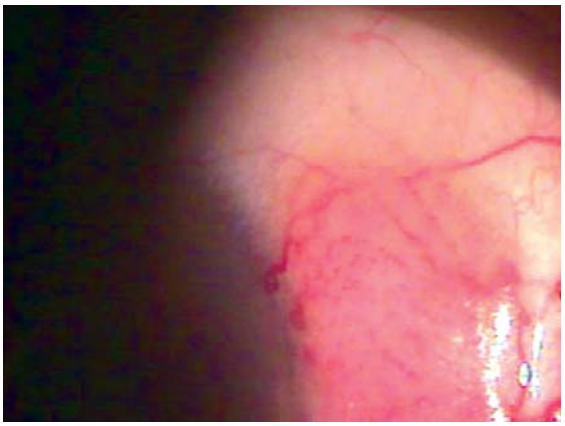

Fig. 13.3: Nodular scleritis

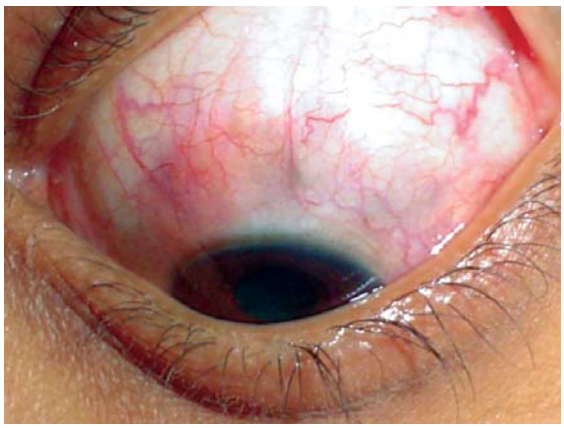

Fig.13.4A: Diffuse anterior scleritis (*Courtesy*: Prof. Manoj Shukla, and Dr Prashant Shukla, AMUIO, Aligarh)

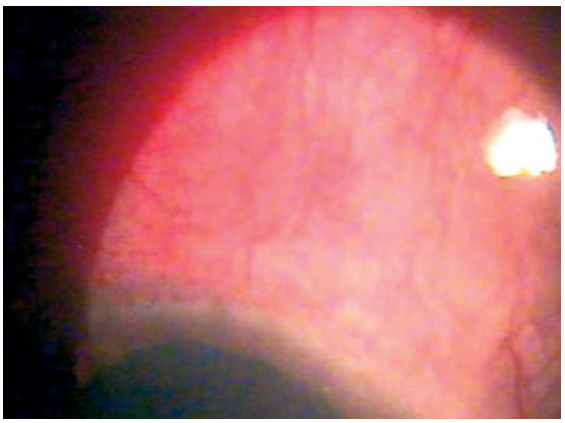

Fig. 13.4B: Nonnecrotizing acute diffuse anterior scleritis

Anterior Necrotizing Scleritis

Anterior necrotizing or brawny scleritis is an acute and serious form of scleritis characterized by intractable pain and violent inflammatory reaction in a localized part of the sclera. There may be an area of local infarction owing to the occlusive vasculitis. The inflammation spreads to the adjoining areas. It often results in destruction of the tissue. The condition leads to anterior uveitis, and may involve the entire anterior sclera causing thinning and subsequent ectasia. Severe visual loss is not uncommon.

Posterior Scleritis

The posterior scleritis is a rare type of scleritis which poses difficulty in diagnosis. The disease is often unilateral and causes pain, diplopia and diminution of vision. In the presence of limitation of ocular movements, proptosis, papilledema and exudative detachment of the retina, the disease should be suspected. Presence of thickened posterior sclera on CT scan or MRI may be helpful in the diagnosis.

Complications Complications of scleritis include selerokeratitis (37%), scleral thinning (33%), uveitis (30%), glaucoma (18%), and cataract (7%).

Treatment Mild cases of diffuse and nodular anterior scleritis respond to oral NSAIDs. Two NSAIDs may be tried in succession in case of therapeutic failure. If NSAIDs are ineffective in managing the inflammation, systemic corticosteroids should be added. Topical therapy is generally ineffective but prednisolone and cycloplegic eye drops are recommended to prevent uveitis. Necrotizing scleritis almost always needs oral corticosteroids. The use of sub-Tenon depot steroids may cause scleral necrosis, hence is not used. If scleritis is not responding to oral corticosteroids, systemic immunosuppressive agents such as methotrexate, cyclophosphamide or cyclosporine, are recommended.

Microbial Scleritis

Bacterial or fungal scleritis is not common. It may occur following trauma by contaminated foreign body and pterygium excision with mitomycin C application. A suppurative scleritis can threaten the eye. Systemic antimicrobial treatment is initiated without corticosteroid or immunosuppressive therapy.

Necrotizing Scleritis without Inflammation (Scleromalacia Perforans)

Scleromalacia perforans (Fig. 13.5) name is a misnomer because it occurs due to inflammation. However, the typical signs of inflammation such as pain and redness do not manifest. It is a rare entity characterized by thinning and melting of the sclera and development of holes without any evidence of scleritis. The disease is common in elderly females usually suffering with severe rheumatoid arthritis. Fibrinoid necrosis of the sclera occurs with exposure of the uveal tissue unassociated with painful symptoms. The damaged sclera may bulge. Trivial trauma may lead to perforation of the globe.

Corticosteroid therapy is contraindicated as there is a danger of impending perforation. The ectatic areas should be repaired by scleral grafting.

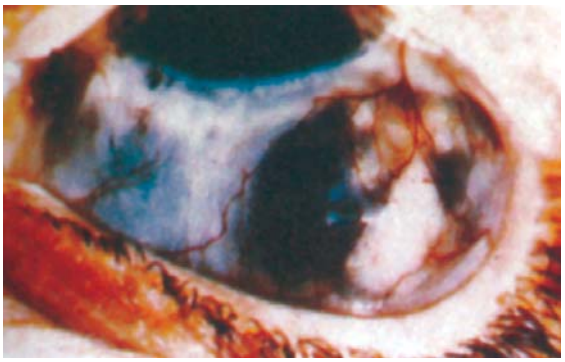

Fig. 13.5: Anterior necrotizing scleritis with scleral thinning and ectasia (*Courtesy*: Sankara Nethralaya, Chennai)

STAPHYLOMAS

Staphyloma is defined as an ectatic cicatrix of the cornea or the sclera in which the uveal tissue is incarcerated. It occurs due to weakening of the outer tunic of eye by an inflammatory or degenerative condition. Trauma and sustained increase in the intraocular pressure are the other contributory factors. Anatomically, staphyloma is classified into following five categories:

1. Anterior (corneal)
2. Intercalary
3. Ciliary
4. Equatorial, and
5. Posterior.

Anterior Staphyloma

After the perforation of a large sloughing corneal ulcer, a pseudocornea may be formed. It mainly consists of organized exudates and fibrous tissue lined anteriorly by the epithelium and posteriorly by the iris. It tends to become ectatic and is called *anterior staphyloma*. It may be partial (Fig. 13.6) or total. There is no anterior chamber but posterior chamber becomes deep.

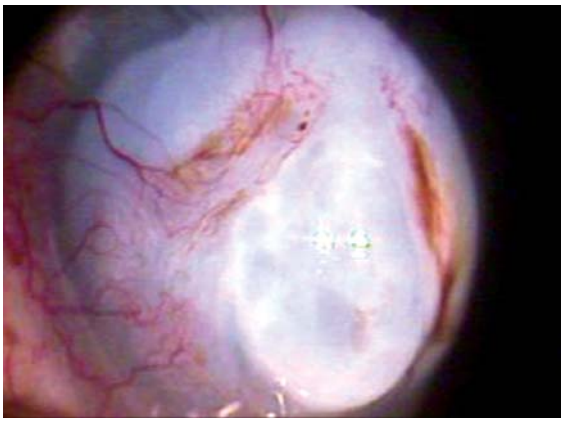

Fig.13.6: Partial anterior staphyloma

Intercalary Staphyloma

Intercalary staphyloma is a localized ectasia of limbal tissue lined by the root of the iris (Fig. 13.7). Perforating injury of the peripheral cornea or perforation of marginal corneal ulcer are implicated in the etiology. The staphyloma may cause visual disturbances due to astigmatic error. There is always a possibility of rise of intraocular pressure owing to extensive peripheral anterior synechiae formation. Localized staphylectomy and iridectomy may prevent the secondary glaucoma.

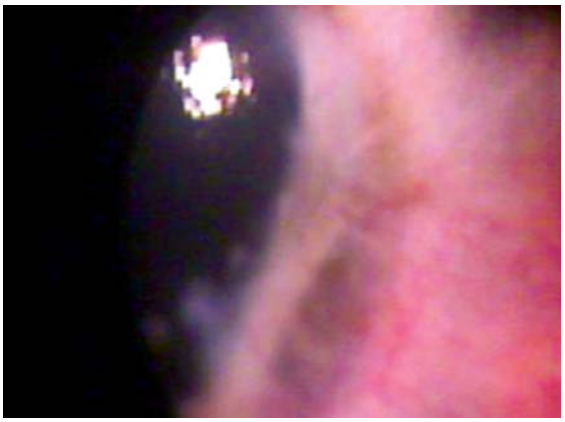

Fig. 13.7: Intercalary staphyloma

Ciliary Staphyloma

Ciliary staphyloma (Fig. 13.8) occurs in the ciliary zone which extends about 8 mm from the limbus. A portion of the ciliary body incarcerates in the

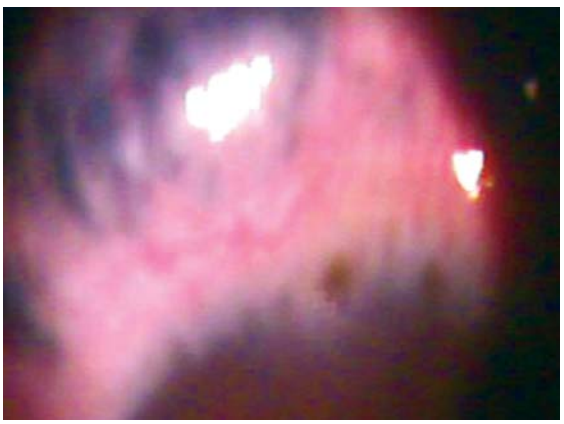

Fig. 13.8: Ciliary staphyloma

ectatic sclera. It looks bluish in color and may be lobulated. Scleritis, trauma to the ciliary region, congenital glaucoma and absolute glaucoma are the common causes of ciliary staphyloma. The ciliary staphyloma may increase in size if intraocular pressure remains elevated.

Equatorial Staphyloma

The ectasia of the sclera of equatorial region with incarceration of the choroid is known as *equatorial staphyloma*. The sclera in the equatorial region is relatively weak due to the passage of four venae vorticosae. Degenerative changes in high myopia and following recurrent inflammatory episodes of scleritis may further weaken the sclera ultimately causing ectasia. It is often found in eyes with absolute glaucoma.

Posterior Staphyloma

Posterior staphyloma is an ectasia of the sclera at the posterior pole which is lined by the choroid. It is found in high degree of axial myopia primarily due to the degeneration of posterior ocular coats. The ectasia presents a crescentic shadow 2 to 3 disk diameters to the temporal side of the optic disk on indirect ophthalmoscopy. The retinal vessels change their course in the ectatic region.

Treatment Prompt treatment of scleritis and control of raised intraocular pressure may prevent staphyloma formation in large number of cases. Localized staphylomas can be repaired by scleral grafting. Staphylectomy or enucleation may be performed in a blind disfiguring eye.

Blue Sclera

The sclera in babies appears blue due to shining of the underlying uveal tissue as the scleral collagen fibers are thin and immature. Blue discoloration of the sclera is pronounced in osteogenesis imperfecta, Ehlers-Danlos syndrome, pseudoxanthoma elasticum and Marfan's syndrome. It may also be found in association with keratoconus and keratoglobus.

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CHAPTER

14 Diseases of the Uveal Tract

ANATOMY

The middle vascular tunic of the eye comprising iris, ciliary body and choroid is called the *uveal tract*.

Iris

The iris is a delicate diaphragm placed between the cornea and the lens (Fig. 14.1). It has a circular opening of about 4 mm called the *pupil* lying eccentrically slightly towards the nasal side. The iris is attached at its periphery (root) to the middle of the anterior surface of the ciliary body.

The root is the thinnest part of the iris and, hence, prone to tear on trauma. The thickest part of the iris is at the collarette which lies about 1.5 mm from the pupillary border. The collarette divides the iris into the pupillary and the ciliary zone (Fig. 14.1). The pupillary margin rests on the anterior surface of the lens. When the lens is absent it becomes tremulous. The pupillary margin appears dark due to anterior termination of the pigment layer of the iris. On either side of the collarette there are several dark pits or crypts passing into the stroma.

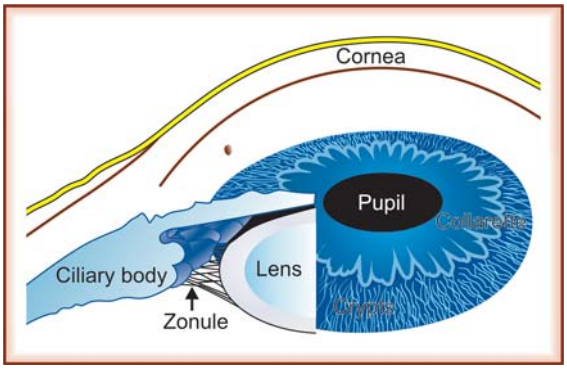

Fig. 14.1: Gross anatomy of the iris

Microscopically, the iris is composed of four layers anteroposteriorly. The anterior limiting layer is a condensation of the anterior part of the stroma and consists of a sheet of fibroblasts. The iris stroma is a loosely arranged collagenous network in which the sphincter pupillae muscle, vessels and nerves of the iris and pigment cells are embedded. The anterior epithelium is essentially a layer of nonstriated muscle cells, the dilator pupillae. The posterior pigment epithelium is a forward extension of the ciliary epithelium and is densely pigmented.

Ciliary Body

The ciliary body is roughly triangular in cross-section with base forwards (Fig. 14.2). It extends anteriorly to the scleral spur (1.5 mm posterior to the limbus) and posteriorly as far as the ora serrata. The ciliary body is composed of unstriated ciliary muscle fibers, stroma and blood vessels. The inner surface of the ciliary body has two distinct zones. The anterior is corrugated with 70 to 80 ridges and is called *pars plicata*. The posterior zone (about 4 mm) is smooth and is known as *pars plana*.

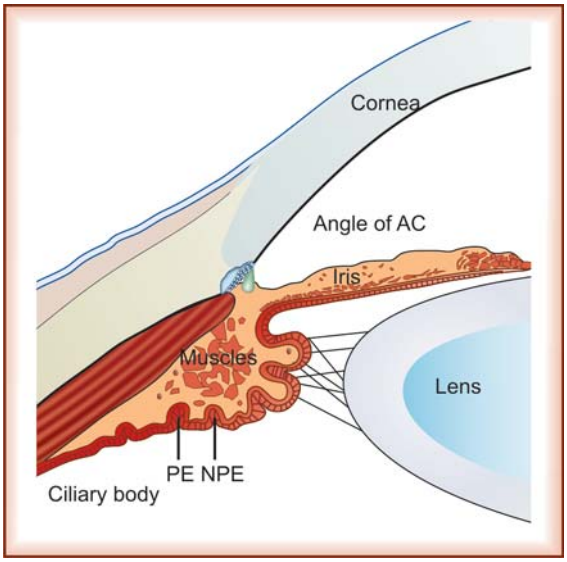

Fig. 14.2: Gross anatomy of the ciliary body.
PE: Pigmented epithelium, NPE: Nonpigmented epithelium

Histologically, the ciliary body has, from without inwards, four layers: suprachoroidal lamina, the ciliary muscle and stroma, the epithelium and the internal limiting membrane (Fig. 14.3). The suprachoroidal lamina consists of pigmented collagen fibers.

The ciliary muscle is mainly composed of three distinct types of fibers: (i) meridional or longitudinal, (ii) radial or oblique, and (iii) circular. The ciliary muscle plays a dominant role in accommodation and facilitates the drainage of aqueous humour by opening the exit channels at the angle of the anterior chamber.

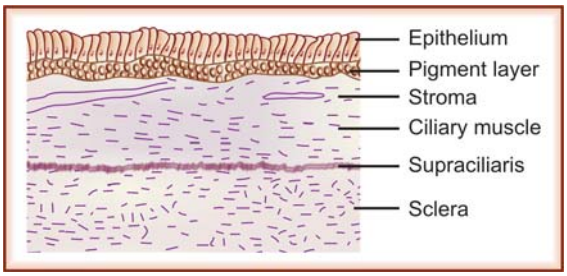

Fig. 14.3: Diagram showing various layers of the ciliary body

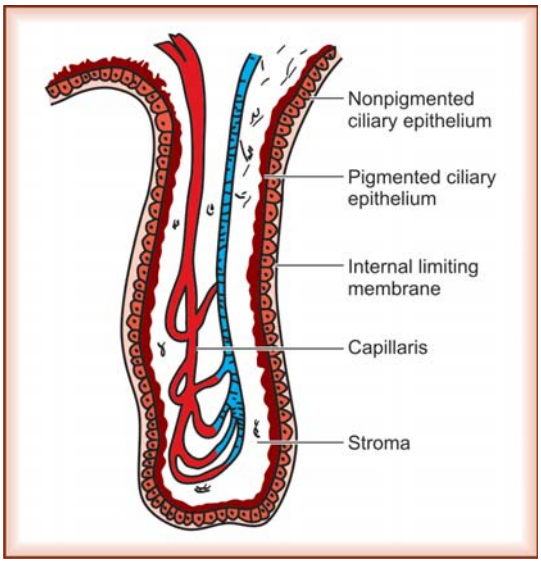

Fig. 14.4: Ciliary process

The ciliary processes (Fig. 14.4) consist essentially of blood vessels, particularly the veins, represent a forward continuation of the choroidal vasculature except choriocapillaris.

The stroma of the ciliary body resembles that of the choroid and is composed chiefly of thick bundles of collagen fibers.

The epithelium is two layered, the outer pigmented and the inner nonpigmented. The nonpigmented epithelium secretes aqueous humor. The internal limiting membrane lines the nonpigmented epithelium and is a forward continuation of the internal limiting membrane of the retina.

Choroid

The choroid is a thin vascular membrane extending from the optic disk to the ora serrata. It is largely composed of layers of large and small vessels (Fig. 14.5). Pigment cells, wandering cells, smooth muscle fibers, and nerves in the intervascular spaces form the stroma of the choroid.

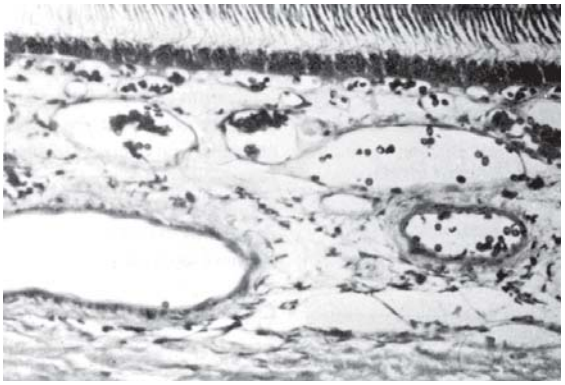

Fig. 14.5: Microphotograph of choroid
(*Courtesy*: Dr M Kincaid, Bethesda Eye Inst. St Louis)

The choriocapillaris are composed of a single layer of endothelial tubes. They nourish the outer part of the retina. The choroidal vessels are bounded by Bruch's membrane internally and suprachoroidal lamina externally.

Blood Supply of the Uveal Tract

The uveal tract is supplied by:

1. Short posterior ciliary arteries
2. Long posterior ciliary arteries, and
3. Anterior ciliary arteries.

The short posterior ciliary arteries supply the choroid. The long posterior ciliary arteries and the anterior ciliary arteries supply the iris and the ciliary body. The short posterior ciliary arteries divide into 10 to 20 branches which pierce the eyeball around the optic nerve to supply the choroid (Fig. 14.6). The blood supply of choroid is essentially segmental. Two long posterior ciliary arteries pierce the sclera obliquely on the medial and lateral sides of the optic nerve. They do not branch until they reach the ciliary muscle. These arteries anastomose with each other and with the anterior ciliary arteries to form the *circulus arteriosus iridis major* at the apex of the ciliary body. Several branches from this circle run radially through the iris dividing dendritically and forming loops which anastomose near the pupillary margin to form the *circulus vasculosus iridis minor*.

The venae vorticosae and their anterior tributaries drain the blood from the uveal tract. The

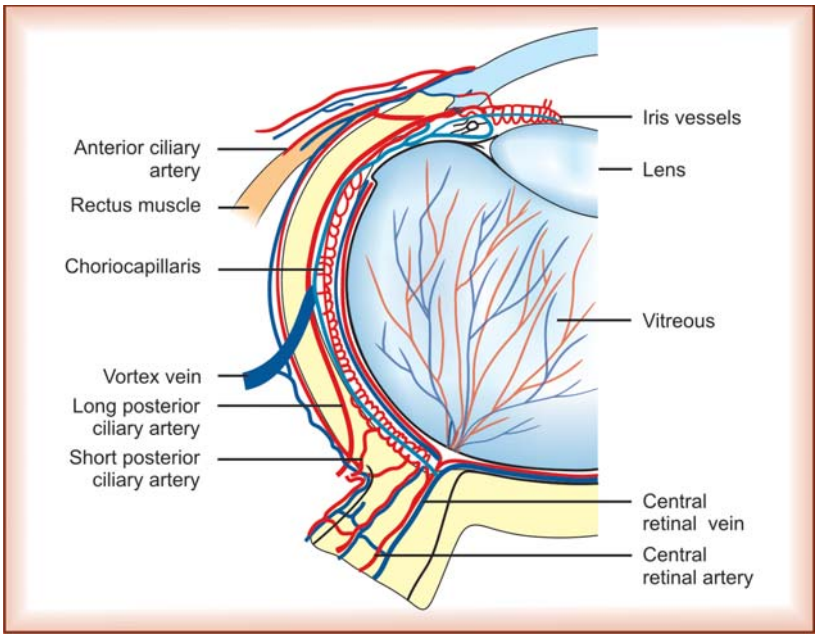

Fig. 14.6: Diagram showing blood supply of uveal tract

anterior ciliary veins carry blood from the outer part of the ciliary muscle.

Nerve Supply of the Uveal Tract

The iris has a dual nerve supply. The parasympathetic nerve fibers arise from the III cranial nerve nucleus in the midbrain, pass along the inferior division of the oculomotor nerve and reach the ciliary ganglion. The postganglionic fibers run in the short ciliary nerves which penetrate the sclera to reach the suprachoroidal space and supply the sphincter pupillae.

The sympathetic fibers reach the dilator pupillae either through the nasociliary branch of the ophthalmic division of the trigeminal nerve via a plexus in the suprachoroid or through the ciliary ganglion.

The sensory supply of the iris is through the nasociliary nerve. The ciliary body derives its sensory supply from the trigeminal nerve through the ciliary nerves, while the motor supply to the ciliary muscle comes from the short ciliary nerves. The choroid is supplied by the branches of the ciliary nerves which are derived from the carotid sympathetic plexus.

DISEASES OF THE UVEAL TRACT

The diseases of the uveal tract may be classified as:

1. Inflammatory
2. Degenerative
3. Congenital, and
4. Neoplastic.

INFLAMMATION OF THE UVEAL TRACT (UVEITIS)

The uveal tract is a vascular membrane, therefore, the inflammatory process tends to affect the uvea as a whole and does not remain confined to a single part. This is especially true for the iris and the ciliary body, hence, the inflammation of the iris (iritis) is almost always accompanied with some inflammatory reaction of the ciliary body (cyclitis) and *vice versa*. Owing to the segmental blood supply of the choroid, the choroidal lesions are often restricted to isolated sectors.

Depending on onset, pathology and etiology, uveitis can be classified in the following ways:

1. **Onset**
   - a. Acute
   - b. Chronic
2. **Pathology**
   - a. Suppurative
   - b. Nonsuppurative
     - i. Nongranulomatous
     - ii. Granulomatous
3. **Etiology**:
   - a. Infectious uveitis
     - i. *Bacterial*
       - Tuberculosis
       - Leprotic
       - Gonococcal
     - ii. *Spirochetal*
       - Syphilis
       - Lyme disease
       - Leptospirosis
     - iii. *Viral*
       - Herpes
       - Cytomegalovirus disease
     - iv. *Fungal*
       - Presumed ocular histoplasmosis syndrome
       - Candidiasis
   - b. Parasitic uveitis
     - Toxoplasmosis
     - Toxocariasis
     - Onchocerciasis
     - Cysticercosis
   - c. Lens-induced uveitis
     - Phacoanaphylactic
     - Phacotoxic
   - d. Uveitis of unknown etiology
     - Pars planitis
     - Fuchs heterochromic cyclitis
     - Glaucomatocyclitic crisis
     - Vogt-Koyanagi-Harada syndrome
     - Sympathetic ophthalmitis

Birdshot retinochoroidopathy
Acute multifocal placoid pigment epitheliopathy
Geographical choroidopathy

e. Uveitis associated with systemic diseases
   i. *Joint disorders*
      Ankylosing spondylitis
      Juvenile rheumatoid arthritis
      Reiter's syndrome
   ii. *Skin disorder*
      Behçet's disease
   iii. *Respiratory disorder*
      Sarcoidosis
   iv. *Gastrointestinal disorder*
f. Uveitis associated with malignancy
g. Uveitis associated with ocular ischaemia
h. Idiopathic uveitis

The International Uveitis Society Group has proposed the following anatomical classification of uveitis:

1. Anterior uveitis: (a) iritis (b) anterior cyclitis (c) iridocyclitis
2. Intermediate uveitis (pars planitis): (a) posterior cyclitis (b) hyalitis (c) basal retinochoroiditis
3. Posterior uveitis: (a) focal, multifocal, diffuse choroiditis (b) chorioretinitis
4. Panuveitis

There are several limitations of the classification of uveitis into granulomatous and nongranulomatous groups. Sarcoidosis, often classified as the classical example of granulomatous uveitis, can have a nongranulomatous presentation also. On the other hand sympathetic ophthalmitis, caused by hypersensitivity to melanin or retinal S-antigen, presents histological features of granulomatous panuveitis. In spite of limitations, the classification is useful in understanding the pathogenesis of the disease.

Sources of Uveal Inflammation

1. Exogenous Sources

Uveitis may occur due to introduction of the infective organism from outside the eye, for example from a penetrating injury or following the perforation of a corneal ulcer.

2. Secondary Sources

Corneal ulcer, deep keratitis, scleritis and retinitis may extend to involve the uveal tract and cause uveitis.

3. Endogenous Sources

The primary infection lies elsewhere in the body such as in teeth, tonsils, lungs, joints and sinuses and reaches the eye through blood. Bacterial, viral, fungal and protozoal infections are identified. As organisms are not demonstrated in all endogenous uveal infections, it is suggested that cellular immunity plays a dominant role in the mechanism of uveitis.

4. Allergic Sources

Allergic uveitis is common and is due to hypersensitivity reaction to the microorganisms or to their proteins and toxins. A latent bacteremia or viremia causes sensitization of the uveal tissue with formation of antibodies, later when there is a renewal of infection the antigen reaches the uvea and results in a severe antigen-antibody reaction.

5. Autoimmune Disorders

Autoimmunity may play a significant role in the pathogenesis of uveal inflammation. The mechanism through which autoimmunity to self-antigens can be triggered is a molecular mimicry. Uveitis is often found in association with rheumatoid arthritis, systemic lupus erythematosus, Wegener's granulomatosis, polyarteritis nodosa, Still's disease (in children), Reiter's disease, Behçet's syndrome

and ankylosing spondylitis, all of which are considered as autoimmune disorders.

HLA and Uveitis

In man there are about 70 different human leukocyte antigens (HLA). Each individual may be characterized by HLA typing which identifies his HLA phenotype. There is a growing evidence to suggest that an association exists between HLA and the disease process which can be genetically determined (Table 14.1). The precise mechanism of disease susceptibility of a normal person who is positive for HLA is still unknown. There are, however, a number of hypotheses; the antigen may act as a favorable receptor site for certain pathogens or the HLA may be linked to the genetic material on an immune response gene, the latter may be responsible for the disease process.

Pathology

Both suppurative and nonsuppurative inflammations occur in the uvea. The nonsuppurative inflammation is usually of two types—nongranulomatous (exudative) and granulomatous (proliferative). Since pathogenic organisms have not been isolated in nongranulomatous lesion, it is considered to be a hypersensitivity phenomenon. The granulomatous uveitis is thought to be due to an invasion of the tissue by causative organisms such as *Mycobacterium tuberculosis* or *Toxoplasma gondii*. However, the organisms are rarely isolated from the lesion. The nongranulomatous uveitis frequently involves the anterior uvea, while the granulomatous has a predilection for the posterior.

Table 14.1: Eye diseases with positive HLA types

Disease	*Antigen*
Ankylosing spondylitis	HLA-B27
Behçet's disease	HLA-B5
Herpetic keratitis	HLA-B5
Intermediate uveitis	HLA-DK15
Birdshot retinochoroidopathy	HLA-A29
Presumed ocular histoplasmosis syndrome	HLA-B9, HLA-DR2
Retinal vasculitis	HLA-B44
Ocular pemphigoid	HLA-B12
Reiter's disease	HLA-B27
Vogt-Koyanagi-Harada syndrome	HLA-BW22
Sympathetic ophthalmitis	HLA-DR4
Sarcoidosis	HLA-B8

Nongranulomatous Uveitis

The nongranulomatous uveitis is characterized by an acute onset, short duration and presence of cells and flare in the anterior chamber. It is marked by edema of the uveal tissue, especially anterior uvea, enormous dilatation of the blood vessels and profuse pouring of lymphocytes, plasma cells and fibrin in the anterior and the posterior chamber.

The increased permeability of uveal vessels causes protein transudation from the iris and the ciliary body. The proteineous influx leads to *aqueous flare* in the anterior chamber which appears as suspended dust particles and can be seen by a narrow 2 × 1 mm beam of slit-lamp. Depending on the amount and nature, the aqueous flare can be graded from 0 to 4+ (Table 14.2).

Besides flare, presence of circulating cells is a strong indication of an active inflammation of the uvea. The aqueous cells in the anterior chamber can also be graded on the basis of number of cells seen in a 2 × 1 mm beam of the slit-lamp (Table 14.2). The lymphocytes in the anterior chamber float due to the convection currents of the aqueous humor and adhere to the posterior surface of the cornea and are known as *keratic precipitates* (KPs). The keratic precipitates, a collection of inflammatory cells on the corneal endothelium, are diagnostic of uveitis. Newly formed KPs appear as small, round, white shining dots (Fig. 14.7). Old KPs look dull, crenated and

Table 14.2: Grading of aqueous flare and cells in the anterior chamber

Grading	*0*	*Trace*	*1+*	*2+*	*3+*	*4+*
Aqueous flare	Absent	Very faint	Faint	Moderate: Iris details seen	Marked: Iris details not seen	Intense: Fibrinous aqueous
No. of cells	No cells	< 5 cells	5-10 cells	10-20 cells	20-30 cells	>50 cells

Fig. 14.7: Multiple lymphocytic KPs

Fig. 14.8: Old KPs

pigmented (Fig. 14.8). The KPs originate from the fixed cells of ciliary body. The cells can also migrate to the anterior vitreous.

The uveal tissue is also infiltrated by lymphocytic cells. The edema or water-logging of the iris causes constriction of the pupil which is exaggerated by a dominant activity of the sphincter pupillae. The accumulation of fibrin between the posterior surface of the iris and anterior surface of the lens facilitates the formation of thin *posterior synechiae*, while its presence on the anterior surface of the iris results in filling of the crypts, giving a dull muddy appearance to the iris.

Granulomatous Uveitis

On the contrary, the granulomatous uveitis is a moderate proliferative reaction wherein the entire uveal tissue is grossly infiltrated by epithelioid cells, giant cells and lymphocytes. When the virulence of the offending organism is less and the body resistance good, the cellular aggregation is localized in one or more regions forming nodules. When the nodules develop at the pupillary border they are known as *Koeppe's nodules* (Fig. 14.9). *Busacca's nodules* appear on the surface of the iris and *Berlin's nodules* appear in the angle of anterior chamber.

In granulomatous uveitis, the aqueous flare is minimal, the KPs are macrophagic and posterior synechiae are heavy owing to massive infiltration of the tissue. Large, yellowish, greasy KPs are known as *mutton-fat KPs* (Figs 14.10A and B), that tend to get distributed in a triangular zone inferiorly on the corneal endothelium (Arlt's triangle). Old KPs become crenated, brown or

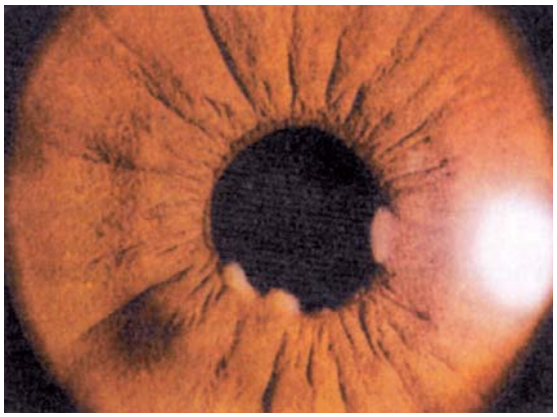

Fig.14.9: Koeppe's nodules

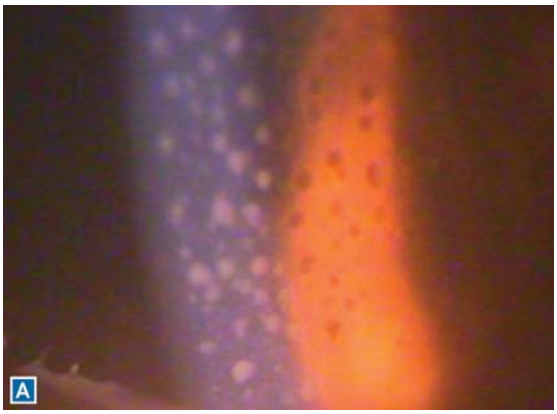

Fig. 14.10A: Mutton-fat KPs

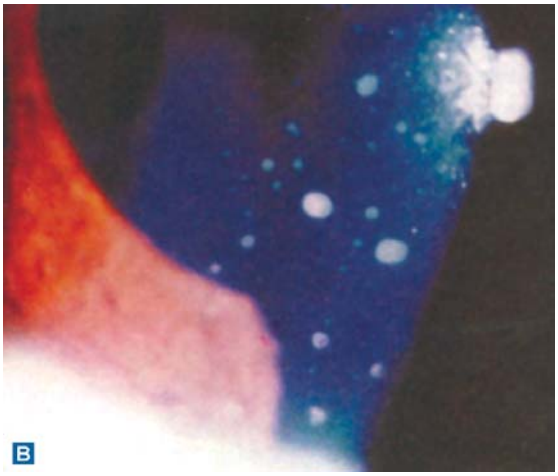

Fig. 14.10B: Mutton-fat KPs under high magnification (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

Table 14.3: Distinguishing features between non-granulomatous and granulomatous uveitis

Features	*Nongranulo-matous*	*Granulomatous*
Onset	Acute	Insidious
Photophobia	Marked	Slight
Visual impairment	Moderate	Marked
Pain	Marked	Minimal
Site	Usually anterior uvea	Usually posterior uvea
Ciliary injection	Marked	Mild
Keratic precipitates	Fine, white, lymphocytic, multiple	Large, gray, macrophagic, few in number
Aqueous flare	+++ (Flare predominates)	++ (Cells predominate)
Iris nodules	Absent	May be present
Posterior synechiae	Thin and weak	Thick and heavy
Vitreous haze	Slight	Moderate to marked

glassy hyalinized and represent resolved uveitis. Chronic or recurrent uveitis may cause iris atrophy, vascularization of the iris and, occasionally, ocular hypotonia following degeneration of the ciliary epithelium.

The differentiation between nongranulomatous and granulomatous uveitis can be made on the points listed in Table 14.3.

Anterior Uveitis (Iridocyclitis)

Clinically, anterior uveitis may manifest in two forms: acute anterior uveitis and chronic anterior uveitis.

Acute Anterior Uveitis

Acute anterior uveitis (Fig. 14.11) is characterized by photophobia, pain, diminution of vision, ciliary injection, presence of fine KPs and aqueous flare, muddy iris, constricted irregular pupil and ciliary tenderness.

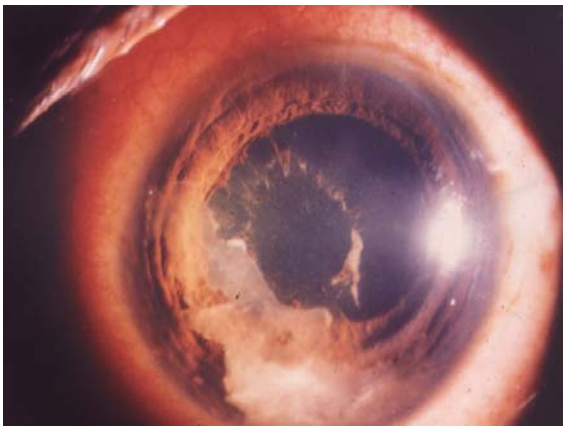

Fig. 14.11: Acute anterior uveitis with fibrin deposit (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

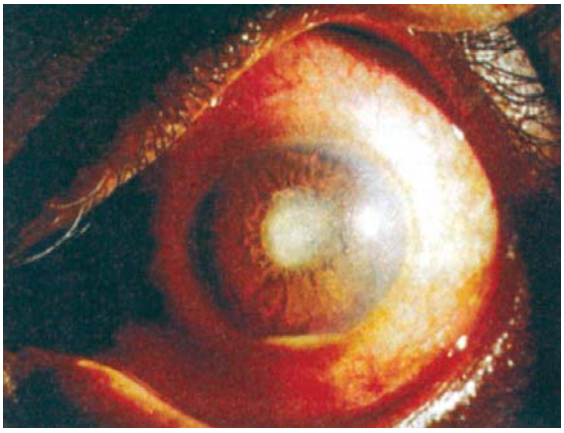

Fig. 14.12: Acute anterior uveitis with hypopyon (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

Clinical features Acute anterior uveitis has a sudden onset with ocular pain usually worst at night. The neuralgic pain often radiates to the forehead, scalp and cheek. The patient complains of photophobia and lacrimation owing to reflex irritation. The vision is slightly blurred in the early phase due to turbidity of the aqueous humor, but marked deterioration of visual acuity may occur in the late stages because of pupillary block by exudates, ciliary spasm, vitreous opacities and cyclitic membrane.

The circumcorneal injection (ciliary flush) or diffuse injection of episcleral vessels is striking. Multiple lymphocytic KPs are present on the back surface of the cornea. Owing to the increased permeability of iris vessels, a moderate to severe reaction may occur in the anterior chamber. The proteinaceous influx leads to aqueous flare while accumulation of polymorphonuclear cells causes hypopyon (Fig. 14.12). Occasionally, erythrocytes get mixed with hypopyon causing a sanguinoid reaction. In herpes zoster and gonococcal anterior uveitis hyphema may be found.

The iris pattern gets blurred and indistinct and the iris appears muddy due to fibrin deposition. The iris becomes edematous and its color fades as compared to the contralateral iris in unilateral uveitis. The pupil is constricted and its reaction becomes sluggish. The eye is tender. Increased viscosity of the plasmoid aqueous and blockage of trabecular meshwork by inflammatory cells cause an elevated ocular pressure (*hypertensive anterior uveitis*). If exudation from the iris and the ciliary body is profuse, it may cover the surface of the iris as well as the pupillary area. This type of uveitis is called *plastic iridocyclitis*.

The exudate facilitates the adhesion of the pupillary margin to the anterior surface of the lens capsule causing posterior synechiae. They are frequently found in the lower part of the pupil due to the gravitational effect of the exudates. The dilatation of the pupil by application of a mydriatic at this stage results in a festooned appearance of the pupil (Fig. 14.13).

Dispersion of pigments on the anterior surface of the lens is almost always found in uveitis. An anterior capsular ring of pigments is often seen in acute iridocyclitis following dilatation of the pupil. It is not rare to find ectropion of the uveal pigment at the pupillary margin consequent to the contraction of organized exudate upon the iris surface.

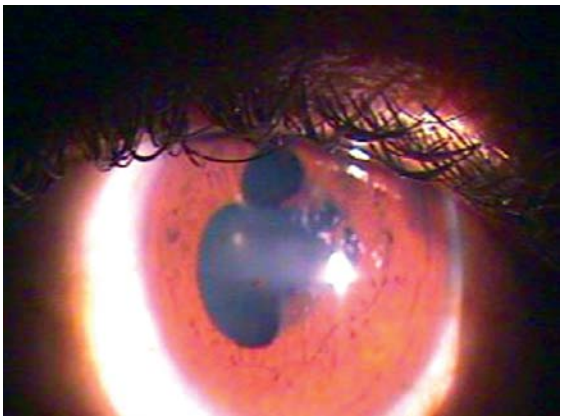

Fig. 14.13: Festooned pupil

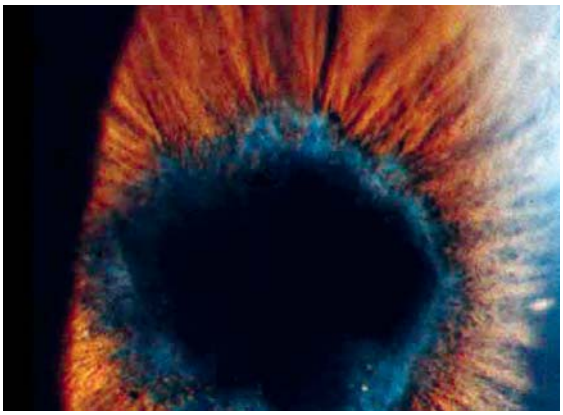

Fig. 14.14: Seclusio pupillae

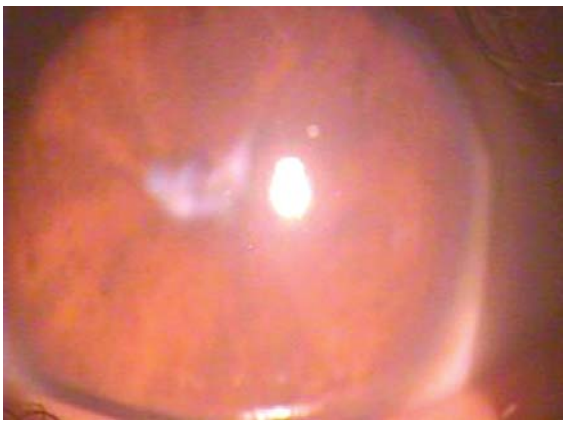

Fig.14.15: Occlusio pupillae

Sometimes, the entire pupillary margin becomes tied down to the lens capsule resulting in the formation of ring or annular synechia or *seclusio pupillae* (Fig. 14.14). The synechia blocks the flow of the aqueous humour from the posterior chamber into the anterior chamber. The aqueous collects behind the iris and pushes the iris forward like a sail, *iris bombé*. The anterior chamber becomes funnel-shaped, deeper in the center and shallower at the periphery. The anterior surface of the iris comes in contact with the posterior surface of the cornea at the periphery where eventually firm adhesions may be formed (peripheral anterior synechiae). Both the ring synechia and the peripheral anterior synechiae may inevitably lead to secondary glaucoma.

Occasionally, the organization of exudates in the pupillary area and the posterior chamber glues the entire posterior surface of the iris to the lens resulting in *occlusio pupillae* (Fig. 14.15) and a total posterior synechia. In this condition, there occurs a retraction of the peripheral part of the iris leading to an abnormally deep anterior chamber at the periphery.

Vitreous involvement in the form of vitritis in acute anterior uveitis is frequent; the inflammatory cells are often found in the anterior vitreous.

Complications

1. **Complicated cataract**: Recurrent iridocyclitis may lead to complicated cataract formation characterized by the presence of polychromatic lustre at the posterior pole when seen on slit-lamp. The cataract rapidly progresses if associated with posterior synechiae. Anterior and posterior subcapsular opacities develop subsequently resulting in a completely opaque lens.
2. **Retrolental membrane**: In severe cases of plastic uveitis, the exudates may form a membrane behind the lens which is known as *retrolental cyclitic membrane*.

Table 14.4: Distinguishing features between acute conjunctivitis, acute anterior uveitis and acute congestive glaucoma

Features	*Acute conjunctivitis*	*Acute anterior uveitis*	*Acute congestive glaucoma*
Colored halos	Present sometimes	Absent	Present
Vision	Normal	Slightly impaired	Grossly impaired
Pain	Little or no pain	Moderate pain in eye along the first division of V nerve	Severe pain in the eye with hemicrania
Discharge	Mucopurulent	Watery	Watery
Injection	Superficial conjunctival	Deep ciliary	Deep ciliary
Anterior chamber depth	Normal	Deep	Very shallow
Iris	Normal	Muddy	Edematous
Pupil	Normal in size and briskly reacting	Small, irregular and sluggishly reacting	Oval, dilated and nonreacting
IOP	Normal	Often normal	Markedly raised
Ciliary tenderness	Absent	Marked	Marked
Media			
a. Cornea	Normal	KPs on the posterior surface	Edematous and hazy
b. Aqueous	Clear	Aqueous flare and cells +	Aqueous flare +
c. Lens	Transparent	Transparent	Cortical opacities may be present
d. Vitreous	Clear	Anterior vitreous hazy	Clear
Constitutional symptoms	Absent	Mild	Prostration and vomiting

3. **Panuveitis and retinal involvement**: The inflammation may extend posteriorly to involve the vitreous and the choroid to produce panuveitis. Cystoid macular edema is not uncommon in longstanding cases of uveitis. Rarely, exudative retinal detachment and neuroretinitis may develop.
4. **Secondary glaucoma**: The rise of intraocular pressure (IOP) is a common complication of acute iridocyclitis. The rise in IOP may occur following clogging of drainage channels by inflammatory cells or debris or by trabeculitis. Pupillary block may cause an acute rise in IOP.
5. **Band-shaped keratopathy**: Longstanding anterior uveitis in children may lead to band-shaped degeneration of the cornea.
6. **Phthisis bulbi**: Recurrent and persistent uveal inflammation causes degenerative changes in the ciliary body. The atrophy of the ciliary epithelium reduces the secretion of aqueous humour resulting in ocular hypotony and eventual shrinkage of the eyeball.

Acute anterior uveitis must be differentiated from acute conjunctivitis and acute congestive glaucoma, the distinguishing features are summarized in Table 14.4.

Chronic Anterior Uveitis

The inflammation of the anterior uvea that lasts longer than three months is termed as *chronic anterior uveitis*. It is characterized by diminution of vision with minimal clinical features of anterior uveitis.

Etiology The etiology of the disease is unknown. It is presumed that chronic iridocyclitis occurs due to a slow release of toxins from septic focus present elsewhere in the body. More commonly the disease is diagnosed during routine examination of the eye.

Clinical features A visual impairment without obvious cause should always arouse the suspicion of the disease. The examination of the eye

reveals mild ciliary injection, tenderness on pressure, scattered or coalesced keratic precipitates on the back of the cornea, a deep anterior chamber and opacities in the vitreous. Sometimes, the presence of KPs on slit-lamp biomicroscopy may be the only sign of the disease.

The disease runs a chronic course and may show exacerbations and remissions. Each attack may further deteriorate the vision and predispose to posterior synechia formation. Repeated attacks may cause iris atrophy and neovascularization associated with an intractable glaucoma. Occasionally, the eye may become soft.

Treatment The anterior uveitis should be treated promptly to prevent complications and sequelae. Both topical and systemic therapies are recommended.

Cycloplegics: The pupil is dilated with atropine sulphate (1% drops or ointment 2 to 3 times a day). The drug gives rest to the eye by paralyzing the ciliary muscle. The dilatation of the pupil prevents the formation of posterior synechia and breaks any if already formed. Atropine diminishes hyperemia but at the same time increases the flow of antibodies by dilatation of the blood vessels. If the patient is sensitive to atropine, homatropine or cyclopentolate (1%) may be used. A more powerful mydriatic effect is obtained by subconjunctival injection of mydricaine (combination of atropine, procaine and adrenaline).

Topical corticosteroids: In acute nongranulomatous anterior uveitis, topical use of corticosteroids (prednisolone, betamethasone or dexamethasone) in the form of suspension or solution gives dramatic results. Initially corticosteroids are used several times a day and when the acute stage subsides, the frequency is reduced. Subconjunctival injections of corticosteroids are helpful in more severe cases of anterior uveitis.

Systemic corticosteroids: Systemic corticosteroids are very effective in nongranulomatous uveitis. Prednisolone 60-80 mg per day (adult dose) is orally administered for 2 weeks and then gradually tapered. The drug has little value in granulomatous anterior uveitis or chronic anterior uveitis.

Antibiotics: Specific antibiotic therapy is often needed. A full course of a broad-spectrum antibiotic is recommended when the cause of the disease remains unknown. Topical antibiotics should be used to prevent secondary infection.

NSAIDs: Topical diclofenic sodium (0.1%) and ketorolac tromethamine (0.5%) are used 3-4 times a day for 4-6 weeks. These drugs are very effective in inhibiting prostaglandin release and act as anti-inflammatory agents. They are safe and do not raise the IOP. Systemic NSAIDs, aspirin, diclofenac or ibuprofen, are useful in relieving the pain.

Immunosuppressive therapy: It may be tried in non-responsive cases of uveitis that are already receiving systemic steroids.

Supportive measures: Hot fomentations are usually soothing and increase the blood-flow and reduce the venous stasis. Dark glasses help in preventing the photophobia.

Management of complications Complications and sequelae of anterior uveitis need energetic and careful management. Hypertensive iridocyclitis requires atropinisation, frequent topical coticosteroid applications and systemic administration of acetazolamide (250 mg, 4 times a day).

Paracentesis is seldom required to control the secondary glaucoma. However, massive hypopyon or hyphema may be managed by paracentesis.

Annular synechia warrants an iridotomy or iridectomy in order to restore communication between the anterior and posterior chambers. The presence of cells in the anterior chamber is usually regarded as a contraindication for intraocular surgery. The extraction of the cataractous lens is

advocated under the umbrella of corticosteroids and antibiotics with guarded visual prognosis. If the eye is phthisical and painful, it should be excised.

Posterior Uveitis (Choroiditis)

The choroidal inflammation may be either nonsuppurative or suppurative. Since the outer layers of retina depend for their nourishment on the choroid, affection of the choroid almost always involves the retina (chorioretinitis).

Nonsuppurative Posterior Uveitis

Nonsuppurative posterior uveitis may be of two types: nongranulomatous and granulomatous.

Nongranulomatous Posterior Uveitis

The nongranulomatous posterior uveitis is also known as *exudative choroiditis* because the inflammatory reaction is marked by exudation and acute leukocytic infiltration in the choroidal layers. It manifests as a white-gray patch with ill-defined edges hiding the choroidal vessels. The patch of choroiditis resolves by fibrosis which appears as a white atrophic area with heaping of pigments at its margin.

Clinical features The symptoms of posterior uveitis are usually visual owing to the involvement of retina and cloudiness of the posterior vitreous. The amount of vitreous haze varies; fine or coarse vitreous opacities and posterior vitreous detachment may be found. The visual deterioration is marked when the lesion lies at the macula. Owing to retinal edema, the images of the objects seen are distorted—metamorphopsia.

The objects frequently appear smaller than their actual size (micropsia) as a result of separation of the rods and cones. Sometimes, the crowding of photoreceptors gives larger images of the objects (macropsia). The patient often complains of flashes of light (photopsia) due to irritability of the retina, and the presence of a black spot before the eye corresponding with the lesion (positive scotoma). Later, subjectively the spot disappears but objectively it can be charted out on visual field recording (negative scotoma). The peripheral choroiditis gives minimum visual symptoms.

Classification The nonsuppurative posterior uveitis is usually bilateral and according to the number and location of the area involved it is classified into the following five sub-types.

1. **Disseminated choroiditis** is syphilitic or tuberculous in origin wherein multiple small lesions are scattered all over the fundus, especially behind the equator (Fig. 14.16). The healed lesions appear as atrophic patches resembling myopic chorioretinal degenerations.
2. **Anterior choroiditis** is often syphilitic and manifests like disseminated choroiditis but it involves mostly the peripheral part of the choroid.
3. **Central choroiditis** (Fig. 14.17) often develops in specific conditions such as toxoplasmosis, histoplasmosis, visceral larva migrans, syphilis and tuberculosis. It may occur in combination with disseminated choroiditis.
4. **Juxtapapillary choroiditis** (Jensen's choroiditis) occurs in young persons and involves the choroid adjacent to the disk, hence the name.

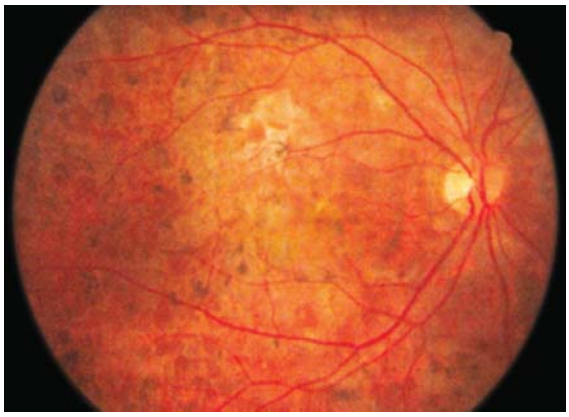

Fig. 14.16: Disseminated choroiditis

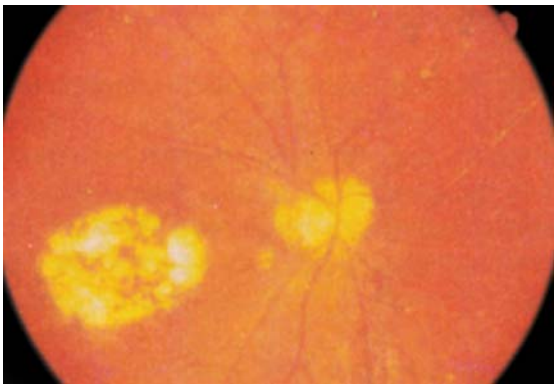

Fig. 14.17: Healed central choroiditis
(*Courtesy*: Dr A Rothova, Donders Institute, Amsterdam)

5. **Diffuse choroiditis** may be caused by tuberculosis or syphilis and is characterized by a raised large yellowish-white or gray plaque with diffuse edges.

Almost all types of choroiditis are associated with exudation in the posterior vitreous causing its haziness. The overlying retina becomes cloudy and retinal vasculitis develops as perivascular cellular cuffing. The choroidal exudates organize and on healing of the disease leave areas of chorioretinal atrophy. Black or slaty pigments heap up on the edges of the atrophic patches over which the retinal vessels course. These must be differentiated from pigments and patches seen in degenerative conditions of the retina, such as pathological myopia and retinitis pigmentosa.

Complications The complications of posterior uveitis include its anterior extension leading to pars planitis or anterior uveitis. A complicated cataract may develop owing to the impairment of the nutrition of the lens. Posterior vitreous detachment and macular edema are common.

Treatment The posterior uveitis is treated on the lines of anterior uveitis. Retrobulbar or periocular injections of corticosteroids are of great help in checking the exudation in the acute phase of disease. Recurrent choroiditis needs sub-Tenon injections of depot corticosteroid.

The systemic administration of antibiotics and corticosteroids often hastens the resolution of the lesion. Once the macula is damaged the visual prognosis becomes poor.

Suppurative or Purulent Uveitis

Panophthalmitis

Suppurative uveitis is characterized by purulent inflammation of the uveal tissue. Although it usually starts as an anterior uveitis or vitritis, it soon involves the whole eye, hence, known as *panophthalmitis*.

Etiology The suppurative inflammation of uvea may occur following penetrating ocular injury, especially with retained intraocular foreign body, and post-operative bacterial or fungal infections. The vitreous is a good culture medium for the growth of pyogenic organisms—*Pneumococcus, Staphylococcus, Pseudomonas pyocyanea, Streptococcus* and *E. coli*. Endogenous panophthalmitis, though rare in occurrence, is metastatic in origin and develops from an infective embolus in the retinal or choroidal vessel.

Pathology Panophthalmitis is marked by polymorphonuclear infiltration into the uveal tissue. A marked tissue necrosis causes the suppurative or purulent exudation in the anterior chamber and vitreous cavity. Cornea, sclera and retina are also involved in the inflammatory process.

Clinical features Panophthalmitis is often accompanied with constitutional symptoms like fever, headache and vomiting. A severe ocular pain occurs associated with marked diminution of vision The eye is proptosed with intense swelling of the lids and chemosis of the conjunctiva (Fig. 14.18). Both ciliary and conjunctival congestions develop. The ocular movements are restricted and painful. The cornea is cloudy and the anterior chamber contains massive hypopyon. The eye is very tender and IOP is often raised.

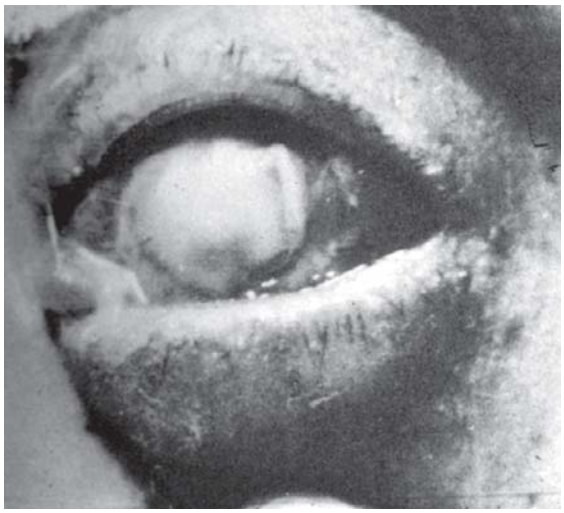

Fig. 14.18: Panophthalmitis

Purulent retinochoroiditis develops and later the vitreous cavity becomes a bag of pus. The posterior lesions cannot be visualized due to haziness of the media. In severe cases, the eyeball may rupture near the limbus, the pus oozes out and ultimately the eye shrinks. Vision is almost invariably lost.

Treatment Panophthalmitis is a serious disease and requires immediate treatment. Perforation of the globe following ocular trauma must be repaired immediately and gentamicin or amikacin be injected subconjunctivally in addition to systemic antibiotics. Postoperative sepsis may be controlled by intravitreal injections of suitable antibiotic and vitrectomy. In desperate cases the eyeball is eviscerated.

Endophthalmitis

Intraocular inflammation involving the vitreous, anterior chamber, retina and choroid is known as *endophthalmitis.*

Types

The endophthalmitis may be classified into two broad groups:

1. Infectious
   i. Exogenous:
      a. Postoperative (0.07-0.12%)
      b. Posttraumatic (2.4-8%)
      c. Bleb infection (0.2-9.6%)
   ii. Endogenous
2. Noninfectious (Sterile).

Infectious Endophthalmitis

Etiology Eyelashes and conjunctiva are the primary source of infection. Intraocular lens (IOL) may act as a vector as bacteria bind to the lens. The microbial endophthalmitis can develop within 1-14 days while fungal endophthalmitis usually develops within three months after injury. *Staphylococcus*, *Propionibacterium* acnes, *Streptococcus*, *Pseudomonas* and *Candida* are common infecting organisms. Nd:YAG laser posterior capsulotomy can precipitate endophthalmitis in specific cases.

Posttraumatic endophthalmitis can develop following penetrating injuries and retained intraocular foreign body.

Microbes can enter the eye through the filtering bleb. Infectious blebitis may develop months or years after the surgery.

Endogenous endophthalmitis results from blood-borne spread of bacteria or fungi during septicemia.

Clinical features Most cases of endophthalmitis have acute onset. Blurred vision, lacrimation, ocular pain and redness are subjective complaints of the patient. Examination of the eye may reveal conjunctival and ciliary injections, chemosis, corneal edema, uveitis, hypopyon (Fig. 14.19), vitritis, tenderness of the eye and hypotonia (occasionally the tension may be elevated). The red fundus reflex may be lost due to vitreal debris or vitreous abscess (Fig. 14.20). Endogenous candida endophthalmitis develops slowly as focal or multifocal areas of chorioretinitis.

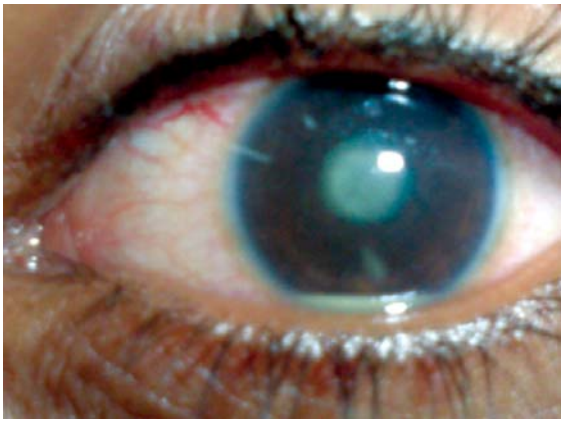

Fig. 14.19: Endophthalmitis with hypopyon (*Courtesy:* Prof. Manoj Shukla, AMUIO, Aligarh)

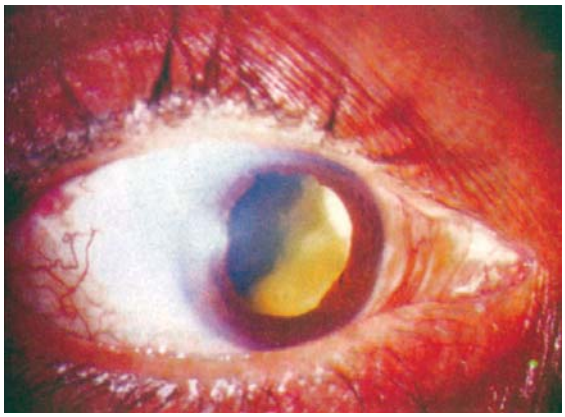

Fig. 14.20: Endophthalmitis with vitreous abscess

Diagnosis Besides complete blood count, blood sugar, serological profile, and X-ray chest examinations are recommended. Aqueous and vitreous taps are obtained and cultured for bacteria and fungi. Aqueous and vitreous smear should be stained by Gram, Giemsa, and calcoflour stains for identification of organisms.

Prophylaxis Preoperative povidone iodine asepsis, topical broad-spectrum antibiotics, preoperative treatment of conjunctivitis, blepharitis, and intraoperative injection of antibiotic may reduce the incidence of endophthalmitis.

Table 14.5: Intravitreal doses of antibiotics and steroid

Drugs	*Intravitreal dose*
Amikacin	0.4 mg/0.1 ml
Amphotericin B	0.005 mg/0.1 ml
Cefazolin	2.25 mg/0.1 ml
Clindamycin	1 mg/0.1 ml
Gentamicin	1 mg/0.1 ml
Vancomycin	1 mg/0.1 ml
Dexamethasone	0.4 mg/0.1 ml

Treatment Broad-spectrum antibiotic coverage by intravitreal route (Table 14.5) for both gram-positive and gram-negative organisms should be provided when etiology is unknown. The Endophthalmitis Vitrectomy Study recommends intravitreal (vancomycin/amikacin), subconjunctival (vancomycin/ceftazidime) and topical (vancomycin/amikacin) antibiotics to treat acute postoperative endophthalmitis.

Patients with severe endophthalmitis are managed by pars plana vitrectomy. The procedure helps in reducing the bacterial load, removing the inflammatory cells, debris and bacterial toxin and clearing the ocular media. The vitrectomy allows better antibiotic penetration following intravitreal injection and provides vitreous for culture and sensitivity testing.

Noninfectious Endophthalmitis

Noninfectious or sterile endophthalmitis is less common and occurs due to retained lens matter or a toxic material introduced during an intraocular surgery. It can be prevented by improving the surgical technique and using good quality intraocular lenses.

Infectious Uveitis

The pattern of uveitis has undergone a sea change. The causes and types of uveitis have been better understood today owing to introduction of more sophisticated methods of investigation. Still a large

number of cases of uveitis remain undiagnosed etiologically while in others the etiology remains presumptive. Some specific types of uveitis are described below.

Bacterial Uveitis

Tuberculous Uveitis

Etiology Mycobacterium tuberculosis can cause either a direct infection or a delayed hypersensitivity reaction in the uvea. Both anterior and posterior uvea can be involved in tuberculosis.

Anterior Uveitis

The involvement of anterior uvea in tuberculosis may occur in three forms—miliary, conglomerate (solitary) and exudative.

1. The *miliary* iritis presents as small grayish translucent tubercles with multiple satellites. The tuberculous nodules may be found in the entire stroma of the anterior uvea but more commonly near the pupillary border. They appear as small gray to grayish-yellow elevations with occasional neovascularization at their base. Hyphema is frequent, and occasionally, pouring of the caseating tuberculous material into the anterior chamber causes pseudohypopyon.
2. *Conglomerate tuberculoma* appears as a yellowish-white granuloma with small satellites seen in young patients. The tuberculoma often erodes the angle of the anterior chamber and causes perforation of the globe.
3. *Acute exudative type of anterior uveitis* with hypopyon, posterior synechiae and vitritis may also occur in tuberculosis.

Posterior Uveitis

Multiple miliary tubercles in choroid (Fig. 14.21) may be found in the terminal stage of tuberculous meningitis. They are seen as round, pale-yellow spots near the optic disk. Diffuse and disseminated choroiditis may be found in chronic tuberculosis. Occasionally, a conglomerate choroidal tuberculoma mimicking a tumor may occur. The accompanying vitreous haze and inflammatory signs in the choroid can distinguish the tuberculoma from the neoplasm.

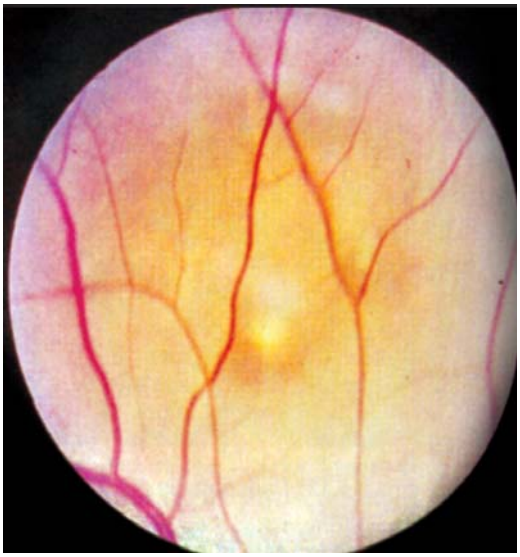

Fig. 14.21: Miliary tubercles in choroid
(*Courtesy*: Dr. J. Biswas, Sankara Nethralaya Chennai)

Complications include retinal vasculitis, dense vitritis, retinal vascular occlusion and papillitis. The extension of tuberculoma may cause perforation of the globe.

Treatment Apart from the usual treatment of anterior uveitis, the antitubercular therapy such as rifampicin and isoniazid must be instituted. Patients on ethambutol need periodical eye examination to prevent toxic amblyopia. In addition to antitubercular treatment, corticosteroids may be necessary in some patients.

Leprotic Uveitis

Etiology Leprosy is caused by *Mycobacterium leprae*. Uveitis is more frequently found in the lepromatous leprosy than in the tuberculoid. There

is an impaired cellular immunity in the lepromatous leprosy, and perhaps, anterior uveitis is a manifestation of antigen-antibody deposition.

Leprosy involves predominantly the anterior uvea. Acute uveitis is usually unilateral, while chronic uveitis is often bilateral and asymmetrical. Its incidence varies from 1 to 42% with a male preponderance (M:F :: 2:1).

Clinical features The anterior uveitis may be either granulomatous or nongranulomatous. The granulomatous anterior uveitis is characterized by the presence of minute yellow pearl-like nodules over the iris without much inflammatory reaction. The non-granulomatous leprotic iritis produces severe exudative reaction.

Treatment In addition to local therapy for anterior uveitis, systemic sulphones must be administered for one to two years. Local and systemic corticosteroids with dapsone (100 mg daily) check the acute inflammatory reaction.

Clofazimine and rifampicin are the other drugs recommended by the WHO for the treatment of leprosy. They are potent drugs effective against the resistant cases. Clofazimine is administered either 50 mg daily or 100 mg on alternate day. Rifampicin is given in a single dose of 600 mg monthly.

Gonorrheal Uveitis

Etiology Gonorrhea is caused by *Neisseria gonorrhoeae* a gram-negative cocci that typically appear in pairs.

Clinical features Gonorrheal uveitis is a metastatic infection almost always affecting males. It is bilateral, the second eye may be affected after some time. Gonorrhea preferentially involves the anterior uvea and the gonococcal anterior uveitis is marked by the presence of gelatinous or greenish-gray hypopyon. Hyphema is common. A less characteristic form of uveitis may occur in gonorrhea associated with arthritis.

Treatment Intensive local corticosteroids and systemic penicillin therapies yield dramatic results. Spectinomycin 2 g twice daily IM for 2 days should be used when the patient is allergic to penicillin. Cefotaxime 1 g IM as one time dose or oral norfloxacin 0.8-1.2 g as a single therapy is also recommended.

Spirochetal Uveitis

Syphilitic Uveitis

Etiology Syphilis is caused by *Treponema pallidum*. It affects both the anterior and the posterior uvea and is capable of producing nongranulomatous as well as granulomatous inflammatory reactions.

Clinical features Bilateral salt-and-pepper fundus, secondary degeneration of retinal pigment epithelium, marked narrowing of the retinal vessels and optic atrophy may be found in congenital syphilis. The fundus picture may mimic retinitis pigmentosa.

Ocular symptoms include pain, photophobia, and blurred vision. A nongranulomatous iritis occurs usually in the secondary stage often accompanied with interstitial keratitis. A gumma may involve the iris or the ciliary body in the secondary or tertiary stage. Gummata are multiple and appear either near the pupillary or ciliary border of the iris. They are yellowish-red in appearance, heavily vascularized and vary in size.

In the early stage of secondary syphilis, a focal or multifocal choroiditis may develop. Exudates may be found around the disk and along the vessels. Other features include retinal vasculitis, exudative retinal detachment, extensive gliosis and pigment proliferation. The condition should be differentiated from retinitis pigmentosa.

Treatment Besides local therapy, systemic administration of penicillin or other antisyphilitic drugs is required.

Lyme Disease

Etiology Lyme disease or borreliosis is a tick-born spirochetal disease caused by *Borrelia burgdorferi*.

Clinical features The course of disease is divided in three stages. *Stage of early infection* is characterized by a distinctive expanding red rash at the site of tick bite (*erythema migrans*). Headache, fever, chills and joint and muscular pain characterize *dissemination stage*. Intermittent joint pain, meningitis, Bell's palsy and cardiac involvement represent *stage of persistent infection*. Ocular involvement occurs in all the three stages. Ocular manifestations of Lyme disease include keratitis, iritis, intermittent uveitis, vitritis, optic neuritis and panophthalmitis.

Diagnosis Lyme immunofluorescent antibody titer, ELISA for IgM and IgG and Western blot test are helpful in the diagnosis of Lyme disease. Serologic testing is not helpful in the diagnosis of early stages of Lyme disease.

Treatment Tetracycline, doxycycline, erythromycin and penicillin therapy is effective in early stage of Lyme disease.

Leptospirosis

Etiology Leptospirosis is caused by *Leptospira* found in secretions from infected animals.

Clinical features Ocular manifestations of the disease include anterior uveitis with or without hypopyon, panuveitis and retinal periphlebitis. Headache, chill, fever, and muscle ache are constitutional symptoms.

Treatment Intravenous penicillin (2.4-3.6 million units per day), tetracycline 500 mg 4 times a day, doxycyclin 100 mg twice a day for 10-14 days may provide relief.

Viral Uveitis

Herpetic Uveitis

Etiology Anterior uveitis may develop in nearly 50% of patients with herpes zoster ophthalmicus, usually manifesting 10-25 days after the onset of the rashes.

Clinical features Typical corneal lesions may be associated with a mild iridocyclitis marked by the presence of KPs, mild flare and moderate cells in the aqueous. A secondary rise of intraocular pressure is not uncommon, it may be associated with atrophy of the iris and damage to sphincter pupillae. Complicated cataract and secondary glaucoma are late complications of the disease.

Treatment Timely institution of topical corticosteroids along with cycloplegic and systemic acyclovir may prevent the complications.

Cytomegalic Inclusion Disease

Cytomegalic inclusion disease (CID) is caused by cytomegalovirus and involves primarily the retina and the posterior uvea, the anterior uvea is affected secondarily. It generally manifests in two forms: *congenital* and *acquired.*

Congenital Cytomegalic Inclusion Disease

Congenital CID affects the neonates. The ocular features include multifocal areas of bush-fire retinochoroiditis in the periphery, anterior uveitis, cataract, hypoplasia of the optic disk and colobomatous microphthalmos. Systemic manifestations include fever, anemia, pneumonitis and hepatosplenomegaly.

Acquired Cytomegalic Inclusion Disease

Acquired CID is often found in acquired immune deficiency syndrome (AIDS) patients. Yellow-white exudates in retina or areas of retinal necrosis,

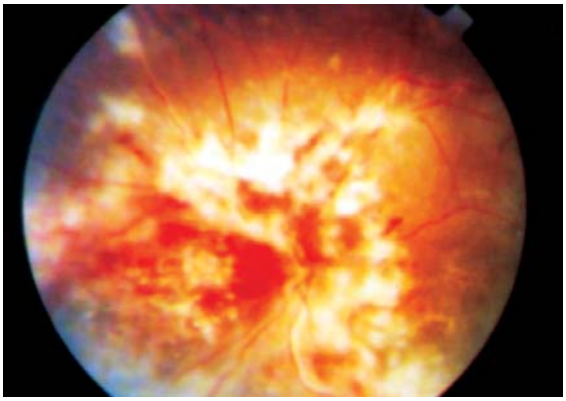

Fig. 14.22: Cytomegalic incision disease retinopathy (*Courtesy:* Dr J Biswas, Sankara Nethralaya, Chennai)

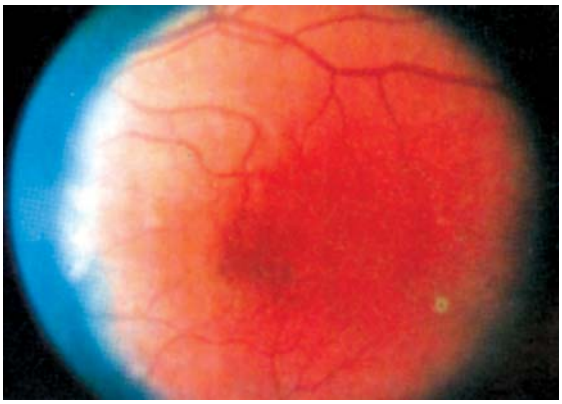

Fig. 14.23: Disciform hemorrhagic retinochoroiditis (*Courtesy*: Prof. Suresh Chandra, Univ. of Wisconsin, Madison)

multiple hemorrhages (Fig. 14.22), vascular sheathing, vitreous exudates and retinal detachment may occur.

Treatment Intravenous ganciclovir (5 mg/kg twice daily for 2 weeks followed by 5 mg/kg once daily long-term maintenance dose), and foscarnet (60 mg/kg 8 hourly for 2 weeks followed by 90 mg/kg daily 5 days/week long-term maintenance dose) have been used with encouraging results. Azidothymidine (AZT) has selective action against human immunodeficiency virus (HIV). A combination of 3 or 4 antiretroviral drugs is known as *highly active anti-retroviral therapy* (HAART) that acts at different stages of HIV life cycle. However, these drugs are toxic to bone marrow and kidney.

Fungal Uveitis

Histoplasmosis

Etiology Histoplasmosis or presumed ocular histoplasmosis syndrome (POHS) is caused by *Histoplasma capsulatum*, a dimorphic fungus with both yeast and filamentous forms. The yeast form is responsible for the ocular and the systemic disease.

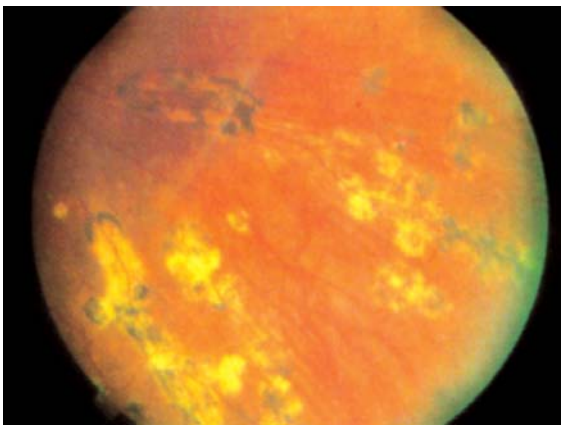

Fig. 14.24: Linear streaks at the equator (*Courtesy*: Prof. Suresh Chandra, Univ. of Wisconsin, Madison)

Clinical features Histoplsmosis is characterized by central or macular disciform hemorrhagic retinochoroiditis (Fig. 14.23), circumpapillary atrophy, punched-out round or oval depigmented small atrophic spots in the peripheral fundus and linear streaks at the equator (Fig. 14.24). The atrophic spots (histospots) probably represent the healed benign histoplasma lesions of childhood. The hemorrhagic disciform maculopathy occurs due to the rupture of Bruch's membrane that results in the development of subretinal choroidal neovascularization. Vitreous cells are not seen in POHS. The patient is usually positive to histoplasmic skin test.

Treatment Amphotericin B, oral and periocular corticosteroids, and destruction of the lesion by photocoagulation have been tried with limited success.

Candidiasis

Etiology Candidiasis is caused by *Candida albicans*. Ocular candidiasis is not common but occasionally occurs in diabetics and patients receiving immunosuppressive therapy or suffering from AIDS.

Clinical features The involvement of posterior uvea by *Candida albicans* is more frequent than the anterior. Anterior uveitis associated with hypopyon may progress to severe panuveitis and vitreous abscess. Multiple white, fluffy, cotton-ball-like lesions of varying size with overlying vitreous haze develop in the choroid. These are associated with retinal hemorrhages and perivascular sheathing.

Treatment Oral flucytosin, fluconazole or rifampin may be administered. Patients with AIDS may need intravenous and intravitreal amphotericin B. Pars plana vitrectomy may salvage the eye.

Parasitic Uveitis

Toxoplasmic Uveitis

Etiology Toxoplasmosis is caused by *Toxoplasma gondii*, a protozoan which primarily involves the central nervous system and retina. *Toxoplasma gondii* have been isolated from the retinal tissue. The parasite causes a granulomatous retinochoroiditis which is typically necrotic.

Toxoplasmosis can be either congenital or acquired.

Congenital Toxoplasmosis

Clinical features The inflammatory reaction is more severe in congenital form than in the acquired

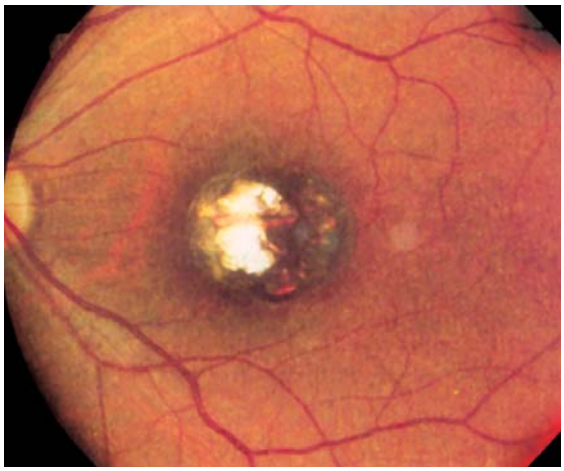

Fig. 14.25: Typical macular lesion of toxoplasmosis

The congenital toxoplasmosis gives a characteristic triad of bilateral punched-out, heavily pigmented macular scars (Fig. 14.25), intracranial calcification and nystagmus.

Floating black spots and blurred vision are common symptoms. The fundus lesion is primarily an exudative focal retinitis and the choroid is only secondarily involved. As the whole thickness of choroid and retina is destroyed in necrotizing inflammation, it leaves a punched-out scars resembling the macular coloboma (Fig. 14.26).

Reactivation of the healed lesion (Fig. 14.27) is quite common and is responsible for significant percentage of posterior uveitis. The recurrence is attributed to the rupture of retinal cyst which releases hundreds of parasites into the unaffected tissue. The fresh lesion appears whitish-yellow and slightly raised commonly occurring at the margin of the old scar.

Punctate outer retinal toxoplasmosis (PORT) is a variant of toxoplasmosis that is seen at the posterior pole with multiple small lesions adjacent to old pigmented scars. These lesions lie at the level of retinal pigment epithelium with minimal vitritis. Rarely, a granulomatous uveitis may develop.

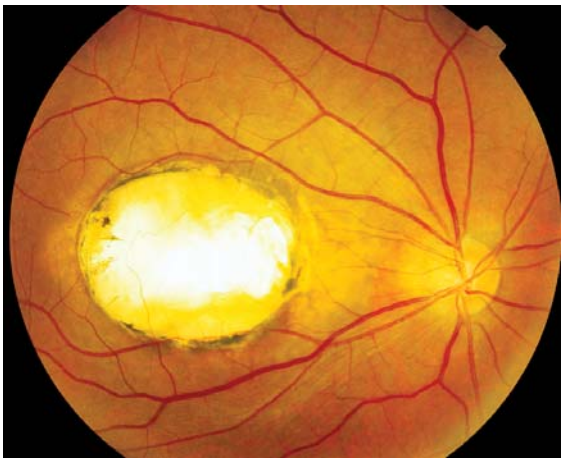

Fig. 14.26: Punched-out macular lesion in toxoplasmosis

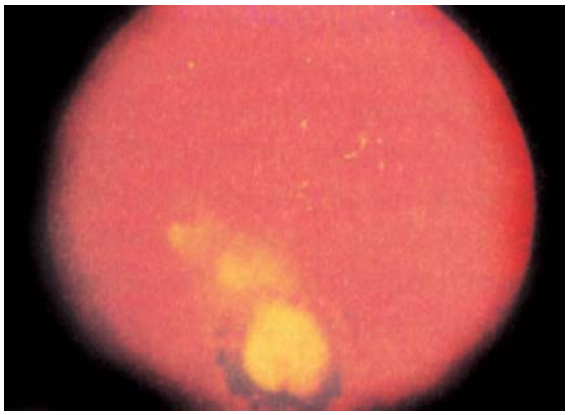

Fig. 14.27: Reactivation of the toxoplasmic lesion (*Courtesy*: Dr A Rothova, Amsterdam)

Acquired Toxoplasmosis

Acquired ocular toxoplasmosis is usually unilateral, mild and without CNS involvement. Some ophthalmologists even doubt its existence.

Diagnosis Sabin-Feldman dye test, enzyme-linked immunosorbent assay (ELISA) for toxoplasma IgG and IgM, differential agglutination test (AC/HS test) using two antigen preparations—AC antigen found in acute infection and HS antigen seen in later stages of infection—and polymerase chain reaction (PCR) amplification to detect *T. gondii* DNA in vitreous, aqueous humor, cerebrospinal fluid, amniotic fluid and blood, are reliable tests for confirming the diagnosis of congenital and acquired toxoplasmosis.

Treatment Pyrimethamine (Daraprim) is administered 25 mg twice a day, after a loading dose of 150 mg, for 5 to 6 weeks. Trimethoprim-sulfamethoxazole can be used as an alternative drug. Sulfatriad 1g four times a day for 6 weeks, and clindamycin 300 g four times a day for 4 weeks are also used. In severe retinochoroiditis associated with vitritis, oral prednisolone (60-100 mg) should be given. Spiramycin is considered as a safe drug and can be combined with sulfadizine for pregnant women. Atovaquone is a cysticidal agent and under investigation. Azithromycin or clarithromycin in combination with pyrimethamine is effective against toxoplasma for short-term treatment.

To prevent leukopenia from pyrimethamine therapy, folinic acid 15 mg thrice weekly should be added. When medical measures fail, photocoagulation and cryotherapy could be used as alternative treatment modalities. Occasionally, pars plana vitrectomy is needed to remove vitreous opacities and membrane.

Toxocariasis

Etiology

Toxocara canis and *Toxocara catis* can cause uveitis in children who play with dogs or cats. Toxocariasis is almost always unilateral and manifests into four clinical forms:

1. Chronic destructive endophthalmitis
2. Posterior pole granuloma
3. Peripheral granuloma, and
4. Vitreoretinal abscess.

Clinical features The child may be asymptomatic or presents with minimal redness, photophobia or strabismus. Granuloma of optic nerve or retina, anterior uveitis, vitritis or neuroretinitis are

commonly seen in toxocariasis. Chronic destructive endophthalmitis is characterized by panuveitis, vitreous clouding and cyclitic membrane formation. It causes severe visual damage and mimics retinoblastoma owing to the presence of leukocoria. Toxocariasis can be diagnosed by the ELISA test.

Treatment Systemic corticosteroids with mebendazole and pars plana vitrectomy may prevent permanent visual loss.

Onchocerciasis

Etiology Onchocerciasis or river blindness is caused by *Onchocerca volvulus*, a filarial nematode, which is transmitted by blackflies of the genus Simulium. It is a major cause of blindness in Central Africa and Southern America.The microfilariae invade the eye and the dead ones induce destructive inflammation.

Clinical features The ocular features are conjunctivitis, snow-flake corneal opacities, corneal scars, sclerokeratitis, anterior uveitis, glaucoma, chorioretinitis (resembling primary retinal degeneration) and optic atrophy. The diagnosis is made by biopsy of the skin nodules.

Treatment Invermectin is a very effective drug. In the community treatment of onchocerciasis, it is given at a dose of 150 μg per kg body weight once a year.

Cysticercosis

Etiology The larva of *Taenia solium* is the most common tape worm that involves the eye.

Clinical features The larvae may be found in the vitreous or the subretinal space of infected patient. A severe zonal granulomatous inflammatory reaction develops around the dead larva causing panuveitis. The presence of cysticercus in the eye is diagnostic. ELISA test may show antibodies to Taenia.

Treatment

Praziquintel 50 mg/kg/day and panphotocoagulation can kill the larva.

Lens-Induced Uveitis

Phacoanaphylactic Uveitis

Phacoanaphylactic uveitis is a zonal granulomatous antigenic reaction to the lens proteins (crystallins). It is basically an autosensitization to the lenticular proteins (antigen).

Etiology Phacoanaphylactic uveitis may develop following disruption of lens capsule after injury, or due to incomplete cortical irrigation and aspiration in extracapsular lens extraction. However, with the advent of modern microsurgical techniques, the incidence of phacoanaphylaxis has decreased dramatically.

Phacoanaphylactic uveitis presents a characteristic microscopic picture. The central necrotic area is composed of lens material infiltrated with polymorphonuclear cells. The pathological picture resembles that of sympathetic ophthalmitis except the necrotic lesion.

Clinical features Clinically, the disease presents as a severe granulomatous anterior uveitis associated with intense pain, marked congestion and blurred vision. Moderate flare and mutton-fat keratic precipitates are often found.

Treatment Topical atropine, and topical and systemic corticosteroids are generally ineffective in the management of phacoanaphylactic uveitis. Removal of all the lens material provides relief.

Phacotoxic Uveitis

Phacotoxic uveitis is a misleading term since there is no firm evidence that the denatured lens proteins are toxic to ocular tissues.

Etiology The phacotoxic uveitis occurs in patients with hypermature cataract (Fig. 14.28). The denatured lens proteins that leak out of the

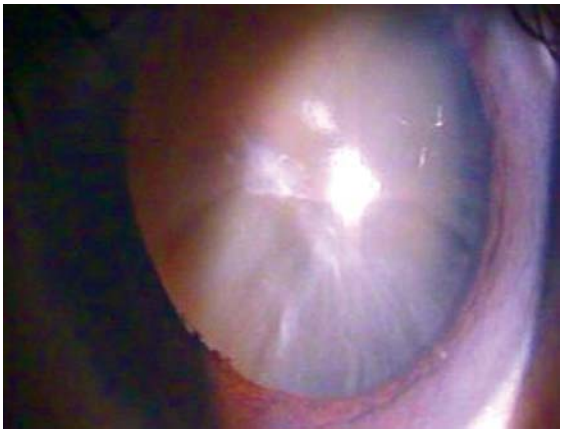

Fig. 14.28: Phacotoxic uveitis

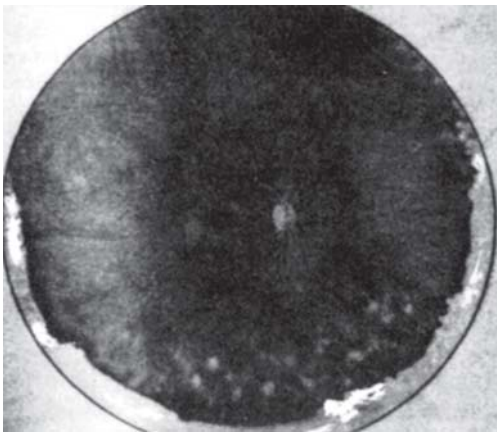

Fig. 14.29: Pars planitis: Snow-ball exudates in lower part of the retina

capsular bag and protein-laden macrophages clog the trabecular meshwork causing severe rise in IOP.

Clinical features Lack of KPs and posterior synechiae, presence of flare and refractile bodies in the aqueous (protein-laden macrophages) and raised IOP are characteristic features. Aqueous tap may show swollen macrophages.

Treatment The management includes prompt cataract extraction following reduction of IOP with osmotic agents.

Uveitis of Unknown Etiology

Pars Planitis

Pars planitis or intermediate uveitis accounts for up to 15% of all cases of uveitis.

Etiology Pars planitis is probably an autoimmune reaction against vitreous, ciliary body and the peripheral retina. An association between pars planitis and HLA-DR2 has been found.

Clinical features Presence of floaters before both eyes is the chief presenting symptom of pars planitis. The anterior segment of the eye is usually quiet. The anterior vitreous contains numerous cells and the indirect ophthalmology reveals snow-ball opacities near the inferior retina (Fig. 14.29). The exudates coalesce to form a white plaque giving a snow-banking effect at pars plana. Peripheral vasculitis associated with sheathing, exudation and vascular occlusion is also common. Occasionally, an anterior segment reaction may be noticed with mild flare and a few KPs. However, posterior synechia is a rare finding. Remissions and exacerbations are seen in 30% cases, while 60% have prolonged course without exacerbation.

Complications Cystoid macular edema may develop in 10-50% of patients with pars planitis. It is a major cause of visual loss. Neovascularization of the retina, vitreous hemorrhages, and tractional or rhegmatogenous retinal detachment may develop. Longstanding cases may present posterior synechiae, vitreous opacities and epiretinal membrane.

Differential diagnosis The differential diagnosis of pars planitis includes sarcoidosis, toxocariasis, syphilis, toxoplasmosis, spillover from iridocyclitis, endogenous endophthalmitis and Lyme disease.

Diagnosis The diagnosis of pars planitis is mainly based on classical clinical features. Laboratory tests are carried out to exclude other causes of intermediate uveitis.

Treatment The four-step approach is used to treat patients with pars planitis:

Step 1: Corticosteroids are considered as first line of treatment. These are injected periocularly by sub-Tenon route every second or third week. If local therapy fails, systemic corticosteroids are administered with an initial dose of 1-1.5 mg/kg/day with gradual tapering. The patient should be maintained on 5-10 mg prednisolone/day. Relapsing cases may need intravitreal triamcinolone injection.

Step 2: If corticosteroids therapy fails, cryoablation or laser photocoagulation of the pars plana is performed.

Step 3: When steps 1 and 2 fail, pars plana vitrectomy with induction of posterior hyaloid separation and photocoagulation to pars plana posterior to the snow-bank may be performed.

Step 4: *When* all the above therapies fail, systemic immunosuppressive agents such as methotrexate, cyclosporin, azathioprine, or cyclophosphamide should be given.

Fuchs Heterochromic Iridocyclitis

Fuchs heterochromic iridocyclitis, a chronic low grade anterior uveitis, is characterized by small, round, diffusely scattered KPs (which never become pigmented), heterochromia of iris and diffuse stromal iris atrophy, and absence of posterior synechia. Cells are often present in the anterior chamber and the anterior vitreous. Neovascularization of the angle of the anterior chamber is usually found. Glaucoma and cataract are common complications. The cataract has good surgical prognosis.

Treatment As posterior synechiae do not develop, cycloplegics are not required. Topical corticosteroids alone may control the inflammation.

Glaucomatocyclitic Crisis

Glaucomatocyclitic crisis or Posner-Schlossman syndrome is marked by intermittent attacks of unilateral secondary open-angle glaucoma associated with low grade iridocyclitis affecting young adults. The attack may last from a few hours to several days.

The patient has minimal symptoms despite the high intraocular pressure (40-60 mm Hg) and the eye is usually white. There may be corneal edema and a few fine nonpigmented keratic precipitates, but no flare and posterior synechia are present.

Treatment Treatment is symptomatic. Topical corticosteroids may provide relief. Timolol maleate and systemic carbonic anhydrase inhibitors are administered to reduce the intraocular pressure.

Vogt-Koyanagi-Harada (VKH) Syndrome

VKH syndrome is an uncommon cause of diffuse uveitis. It often affects persons of Asian ancestry between 30 and 50 years of age. Etiology of VKH syndrome is unknown. An immune reaction to protein associated with uveal melanin may trigger the reaction. HLA-DR4 is strongly associated with VKH syndrome. Histologically, a granulomatous choroidal inflammation resembling with the picture of sympathetic ophthalmitis, but with the distinction of inflammation extending to the choriocapillaris, is seen.

Clinically, the course of VKH syndrome may be divided into 4 phases:

1. *Prodromal phase*: It is marked by neurological symptoms including headache, neck rigidity, hemiparesis, optic neuropathy and tinnitus.
2. *Uveitic phase*: It occurs within 1-2 days after the onset of neurological signs. Photophobia,

blurred vision and redness are usual presentations. It is characterized by the presence of cells in the anterior chamber and vitreous, and exudative retinal detachment in both eyes.

3. *Chronic phase*: It begins with resolution of exudative retinal detachment. Depigmentation of the choroid results in the "sunset-glow" fundus appearance. Multiple small round depigmented lesions in the inferior peripheral fundus are found. Perilimbal vitiligo (Sigiura sign) may be seen in some patients. The extraocular signs include vitiligo, alopecia and poliosis in about 30% of the patients with VKH syndrome.
4. *Recurrence phase*: The disease may reoccur if it is not treated properly. The recurrence is marked by anterior granulomatous uveitis, iris nodules, iris depigmentation and atrophy.

Complications VKH syndrome may lead to cataract (50%), glaucoma (33%) and choroidal neovascularization.

Treatment Cycloplegic agents and corticosteroids must be administered early. Corticosteroids are used topically, periocularly and systemically. Immunosuppressive therapy is considered mandatory in the treatment of VKH syndrome when inflammation cannot be controlled adequately by systemic corticosteroids and/or in patients who develop intolerable adverse effects to steroids.

Sympathetic Ophthalmitis

It is described in the chapter on *Injury to the Eye*.

Retinochoroidopathies

Retinochoroidopathies are inflammatory disorders of unknown etiology involving choroid, choriocapillaris, retinal pigment epithelium and sensory retina. The characteristic features of retinochoroidopathies include single or multiple white, yellow or gray spots in retina without anterior segment inflammation. Later, pigments may be deposited at the periphery of the lesion.

Birdshot Retinochoroidopathy

Birdshot retinochoroidopathy (vitiliginous chorioretinitis) occurs in the fourth decade of life, usually in females. Blurring of vision, color vision defects and nyctalopia are common symptoms.The distribution of spots in the retina resembles the pattern of a birdshot scatter (Fig. 14.30) from a shotgun. The disease may respond to corticosteroid therapy.

Acute Posterior Multifocal Placoid Pigment Epitheliopathy

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) presents multiple cream colored, plaque-like homogenous lesions beneath the retina. The lesions are one disk diameter or less in size but may become confluent. Later, alternate areas of depigmentation and pigment clumping may be found. Vitreous cells and optic disk edema may occur without anterior segment involvement. No effective treatment is

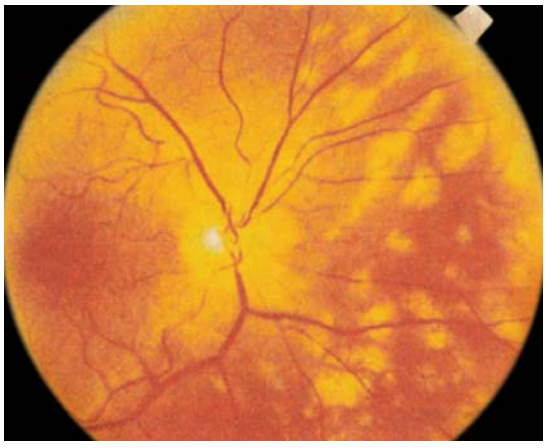

Fig. 14.30: Birdshot retinochoroidopathy
(*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

available. However, most patients recover vision as the fundus lesions run a short self-limited course.

Serpiginous Choroidopathy

Serpiginous choroidopathy or geographical helicoid peripapillary choroidopathy (GHPC) is a bilaterally asymmetrical chorioretinitis marked by the presence of cream-colored irregular patches in the peripapillary region. The lesions spread outwards and leave areas of scarring. Recurrence is the rule. GHPC is a progressive disease, the new lesions spread centrifugally contiguous to the previous scars. The disease may progress relentlessly despite aggressive therapy with corticosteroids and immunosuppressants.

Uveitis Associated with Systemic Diseases

Uveitis Associated with Joint Disorders

Following joint disorders are often associated with acute anterior uveitis and antigen positivity:

1. Ankylosis spondylitis
2. Juvenile rheumatoid arthritis, and
3. Reiter syndrome.

Ankylosing Spondylitis

An acute nongranulomatous anterior uveitis is associated with ankylosing spondylitis in about 15% of cases. It is often unilateral, but the other eye may get involved after some time. The inflammation may last for 2 to 6 weeks. The uveitis is usually recurrent and may lead to macular edema, complicated cataract and glaucoma. The radiograph of sacroiliac region may reveal sclerosis and narrowing of the joint spaces.

Acute anterior uveitis should be managed with topical cycloplegics and corticosteroids. Long-term administration of NSAIDs may help prevent recurrence of the disease.

Juvenile Rheumatoid Arthritis (Still's Disease)

Juvenile rheumatoid arthritis (JRA) is the most common joint disorder associated with chronic iridocyclitis. The disease has 3 types of onset:

1. *Systemic onset* or *Still's disease*: It is found in children below the age of 5-year and have minimal joint involvement. Uveitis is seen in less than 6% of patients.
2. *Polyarticular onset*: Nearly 40% of JRA cases belong to this type involving five or more joints in the first 6 weeks of the disease. Uveitis is found in 7-14% of cases.
3. *Pauciarticular onset*: This type of onset shows involvement of four or fewer joints in the first 6 weeks of the disease. The vast majority (80-90%) of patients with this type of JRA have uveitis.

Clinical features Uveitis occurs within 5-7 years of the onset of joint disease. Moderate pain, photophobia and blurred vision are common symptoms. Eye is often white with fine KPs, cells in the anterior chamber and posterior synechiae.

Complications Complications are frequent and include band keratopathy, cataract, secondary glaucoma, vitreous opacities, macular edema and hypotony.

Treatment Topical short-acting cycloplegics and topical, periocular and systemic corticosteroids should be administered depending on the severity of the disease. To prevent corticosteroid-induced glaucoma, NSAIDs and weekly low doses of methotrexate should be given to control the inflammation.

Reiter's Syndrome

Etiology The etiology of Reiter's syndrome is unknown. It affects mostly young males (16-42 years) positive for HLA-B27 antigen.

Clinical features Nonspecific conjunctivitis, urethritis and polyarthritis characterize Reiter's syndrome. Keratoderma blennorrhagica of palms and soles and balanitis circinata are also found. Usually the patient develops a nongonococcal urethritis which is followed by arthritis, conjunctivitis and anterior uveitis. The conjunctivitis is mucopurulent and may be associated with punctate subepithelial keratitis. An acute nongranulomatous anterior uveitis occurs independently of conjuctivitis in approximately 30% of the cases.

Treatment Reiter's syndrome has a self-limiting course. Besides topical cycloplegics and corticosteroids, tetracycline therapy for 3 to 6 weeks may be effective in chlamydia-induced reactive arthritis. Antiprostaglandins may be helpful in ameliorating the joint swellings.

Uveitis associated with Skin Disorder

Behçet's Syndrome

Behçet's syndrome consists of recurrent uveitis with hypopyon, oro-genital aphthous ulcers and erythema multiforme.

Etiology The etiology of the syndrome is unknown, although the basic lesion is an obliterating vasculitis. The disease occurs more frequently in young adult Japanese.

Clinical Features The classical ocular signs include episodes of acute bilateral nongranulomatous anterior uveitis usually associated with hypopyon. The posterior uveal lesions include focal retinal necrosis, macular edema and ischemic optic neuropathy. Vitritis, periphlebitis retinae and massive retinal exudation can also occur.

The most common skin lesion is erythema nodosum on the legs and ankles. Central nervous system involvement in the form of meningitis, encephalitis and focal neurologic deficits may occur in about 25% of cases.

Treatment Uveitis may not respond to local and systemic corticosteroid therapy. Early use of systemic immunosuppressive agents is likely to improve the long-term visual prognosis.

Uveitis Associated with Respiratory Disorder

Sarcoidosis

Etiology Sarcoidosis is a multisystem granulomatous disease of unknown etiology. It affects the young persons and involves hilar lymph nodes, skin and eyes.

Clinical features Ocular lesions are found in 20 to 50% cases. Uveitis is the most common ocular manifestation of the disease. Chronic granulomatous anterior uveitis (Fig. 14.31) is more characteristic of sarcoidosis though acute anterior uveitis and posterior uveitis can also occur.

The uveitis is bilateral and pleomorphic. Classically, it is granulomatous with minimum symptoms. Mutton-fat keratic precipitates are distributed widely on the entire cornea and do not get collected in a classical inferior triangular manner. Numerous gray-yellow, translucent, vascularized, small or big nodules are found on the iris particularly on the pupillary margin

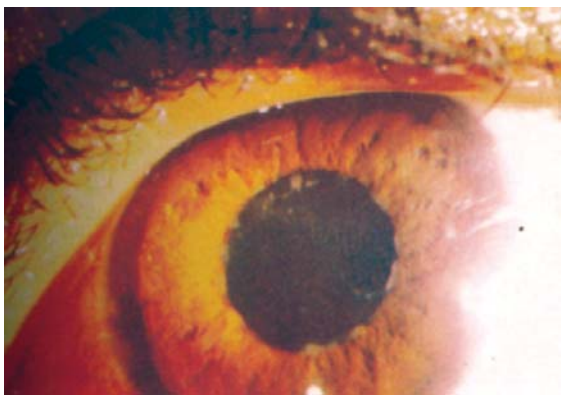

Fig. 14.31: Granulomatous anterior uveitis in sarcoidosis (*Courtesy*: Dr A Rothova, Donders Institute, Amsterdam)

(Koeppe's nodules). When these nodules are seen in the anterior chamber angle (Berlin's nodules) they are characteristic of sarcoidosis. Broad posterior synechia and nummular keratitis may also develop.

Like pars planitis, the vitreous may contain snow-ball opacities overhanging the peripheral inferior retina in sarcoidosis. Nodular granulomas may be found in the retina and choroid. Perivascular candle-wax drippings, retinal periphlebitis, cystoid macular edema, retinal hemorrhages and optic neuropathy are other posterior segment findings seen in sarcoidosis.

Complications Complicated cataract, glaucoma and band-shaped keratopathy are the usual complications. Keratoconjunctivitis sicca and noncaseating granuloma of the lacrimal gland may be found in some patients of sarcoidosis.

Diagnosis Sarcoidosis can be diagnosed by conjunctival, lacrimal gland or lymph node biopsy, raised serum angiotensin converting enzyme (ACE) activity, isotopes studies and radiological evidence of hilar lymphadenopathy. Pulmonary function tests often provide a clue to diagnosis. In the absence of a known etiologic agent, sarcoidosis often remains a diagnosis of exclusion on laboratory and imaging studies. Only tissue biopsy is confirmatory in diagnosing the disease.

Treatment Periocular and systemic corticosteroids and topical cycloplegic drops are the mainstay of therapy of sarcoid uveitis. Antimetabolites such as methotrexate and azathioprine should be administered in non-responding cases.

Uveitis Associated with Gastrointestinal Disorders

Acute uveitis may be found associated with Crohn's disease, ulcerative colitis and Whipple's disease.

Uveitis Associated with Malignancy

An association of uveitis with reticulum cell sarcoma of the brain has been found.

Uveitis Associated with Ocular Ischemia

A low grade ocular ischemia can cause inflammation of the uvea. It alters the permeability of vessels resulting in leakage of cells and proteins.

Idiopathic Uveitis

Nearly 30% of all cases of uveitis are idiopathic.

DEGENERATIVE CHANGES IN THE UVEAL TRACT

Iris

The degenerative changes in the iris are not uncommon.

Depigmentation of iris is associated with atrophy of the iris stroma and found in old people. Iris atrophy may occur as a sequel to anterior uveitis and acute congestive attack of angle-closure glaucoma.

Dehiscence of the anterior mesodermal layers of iris (iridoschisis) may develop as a senile change or may be a late manifestation of ocular trauma. The strands of anterior stroma may float in the anterior chamber.

An *essential atrophy of iris* (Fig. 14.32) is of unknown etiology. It may start in adult life leading to the development of multiple holes in the iris. The atrophy is often unilateral and progressive and affects young females. The atrophy causes more or less complete shrinkage and disappearance of iris tissue and facilitates the formation of peripheral anterior synechiae. The changes at the angle of the anterior chamber can lead to an intractable glaucoma.

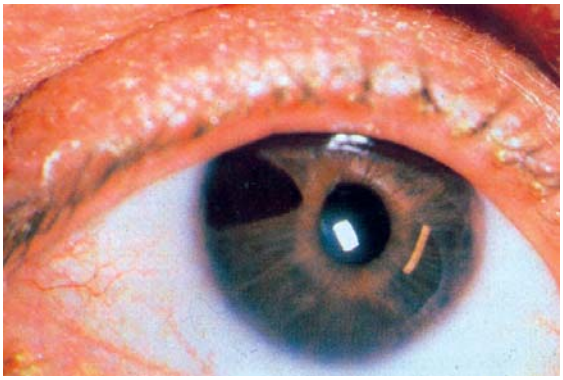

Fig. 14.32: Essential atrophy of iris (*Courtesy*: Dr T Perkins, Madison)

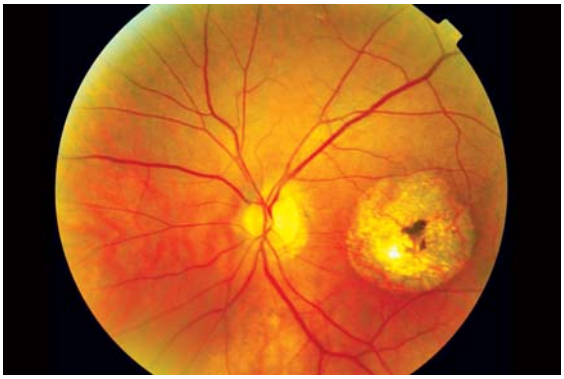

Fig. 14.33: Central areolar choroidal atrophy

Choroid

The degenerative changes in the choroid are more common than in the iris. They may be *primary* or *secondary*. The choroidal changes are common in old age (senile atrophy) and myopia. The changes may be either localized or generalized.

Familial dominant drusen or *central guttate choroidal atrophy* is a bilateral condition marked by the presence of numerous minute, yellowish-white lesions in the macular area (Hutchinson-Tay choroiditis). They are due to the presence of hyaline excrescences on Bruch's membrane known as *colloid bodies*. They generally do not cause visual impairment.

Central areolar choroidal atrophy occurs due to the atrophy of choroid, and is characterized by the appearance of a large circular degenerative patch in the macular area (Fig. 14.33) in which the ribbon-like choroidal vessels are seen.

Gyrate atrophy of choroid is seen in ornithinemia, an inborn error of metabolism, and is characterized by progressive atrophy of choroid and retinal pigment epithelium with macular sparing.

Choroideremia is an X-linked disorder affecting exclusively males. It is characterized by night-blindness, progressive atrophy of the retinal pigment epithelium (RPE) and choroid, and constriction of visual fields. The disease manifests in childhood associated with depigmentation of RPE and progresses to marked atrophy of choroid causing blindness.

Myopic chorioretinal degenerations are commonly seen in pathological myopia. They include myopic crescent, myopic chorioretinal atrophy mimicking choroiditis and Fuchs' fleck. They are described in the chapter on *Errors of Refraction*.

Secondary choroidal degenerations or atrophic changes are quite frequent following chorioretinitis. They are often associated with pigmentary changes.

Detachment of the Choroid

Etiology Separation of choroid from the sclera may occur during the first few days following an intraocular surgery as a result of sudden lowering of intraocular pressure. Severe choroidal hemorrhage, choroidal tumors , intraocular inflammation and trauma are other causes of choroidal detachment.

Clinical Features The anterior chamber becomes shallow, ocular tension is low and a dark brown

mass is seen on funduscopy. Longstanding shallow anterior chamber predisposes to peripheral anterior synechia formation and secondary glaucoma.

Treatment Postoperative choroidal detachment resolves by itself. Oral administration of acetazolamide and drainage of suprachoroidal fluid through a sclerotomy may settle the detached choroid.

CONGENITAL ANOMALIES OF THE UVEAL TRACT

Hetrochromia

The iris shows great variations in its color. When one iris differs in color from the other, the condition is called *heterochromia iridum*. When a sector of iris has a different color from the remainder, it is known as *heterochromia iridis (Fig. 14.34)*.

Anomalies of Pupil

When the pupil is abnormally eccentric it is called *corectopia*. There may be more than one pupil, such a condition is known as *polycoria*.

Aniridia

Aniridia (irideremia) is a rare condition where the iris is absent. Careful examination often reveals the presence of a narrow rim of iris tissue hidden behind the sclera. The zonule of the lens and ciliary processes are often visible. Secondary glaucoma supervenes due to the chamber angle anomalies.

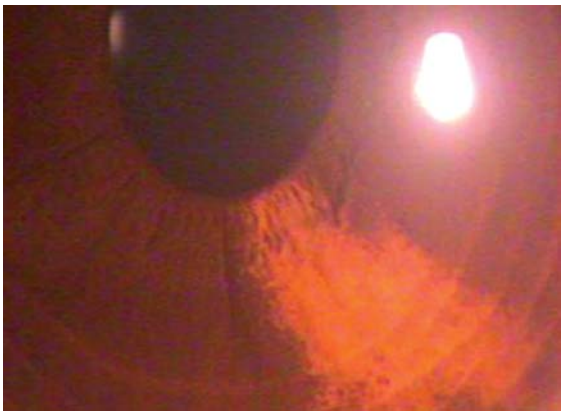

Fig. 14.34: Heterochromia iridis

Persistent Pupillary Membrane

The persistence of a part of the anterior vascular sheath of the lens, which usually disappears, is called *persistent pupillary membrane*. Such remnants are not uncommon in infants, particularly when they are examined on the slit-lamp. The pupillary membrane is usually attached to the collarette. Sometimes, punctate remnants are left on the lens surface. They are small, numerous, stellate-shaped and unassociated with anterior uveitis, and can be distinguished from broken posterior synechiae.

Coloboma of the Uveal Tract

Coloboma of the uveal tract may be typical or atypical.

Typical coloboma of the uveal tract is associated with the nonclosure of the fetal fissure and occurs in the inferior part of the eye. The pupil appears pear-shaped (Fig. 14.35). The choroidal coloboma

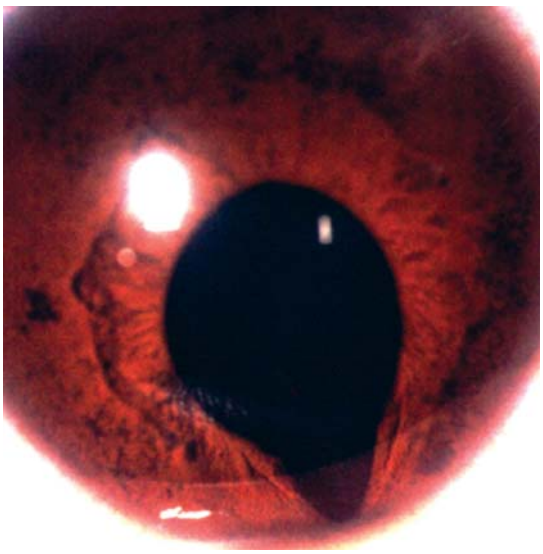

Fig. 14.35: Typical coloboma of iris

is usually oval with a rounded apex towards the disk. A few vessels may traverse over the floor. The coloboma may or may not involve the disk.

Atypical colobomata of the retina and choroid are extremely rare but are relatively common in iris. Intrauterine inflammation and persistence of the fibrovascular sheath of the lens are implicated in the etiology.

Cysts of the Iris

Congenital cysts of the iris may arise either from the stroma or from the pigment epithelium. Stromal cyst is probably derived from the ectopic cells of the surface ectoderm of the developing lens, while the cyst of the neuroepithelium appears due to the failure of fusion of the two layers of optic vesicle.

The congenital cyst of the iris should be differentiated from *implantation cyst of the iris* which occurs after a perforating ocular injury or an intraocular surgery. The implantation cyst has a characteristic pearly appearance. Closure of the iris crypts causes retention of fluid and forms *serous cysts*. *Parasitic cysts* of the iris are rare.

NEOPLASTIC DISORDERS

Tumors of the uveal tract are described in the chapter on *Intraocular Tumors*.

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CHAPTER

15 Glaucoma

The term glaucoma refers to a group of conditions that have a characteristic optic neuropathy associated with visual field defects and elevated intraocular pressure.

Normally the rate of aqueous formation and the rate of aqueous outflow are in a state of dynamic equilibrium and, thus, maintain a normal intraocular pressure which ranges between 12 and 20 mm Hg. Intraocular pressure (IOP) is basically determined by three factors:

1. The rate of aqueous humor production
2. Resistance to aqueous outflow across the trabeculum, especially in the juxtacanalicular meshwork, and
3. The level of episcleral venous pressure.

A brief review of the anatomy of the angle of the anterior chamber is necessary to understand the pathophysiological mechanism of glaucoma.

ANATOMY OF THE ANGLE

The anterior chamber is bounded anteriorly by the posterior surface of the cornea and posteriorly by the anterior surface of the iris and the anterior surface of the lens. It is about 2.5 mm deep in the center and is filled with aqueous humor. The angle of the anterior chamber is a peripheral recess formed by the root of the iris and a part of the ciliary body posteriorly and corneo-sclera (trabecular tissue and scleral spur) anterolaterally (Fig. 15.1).

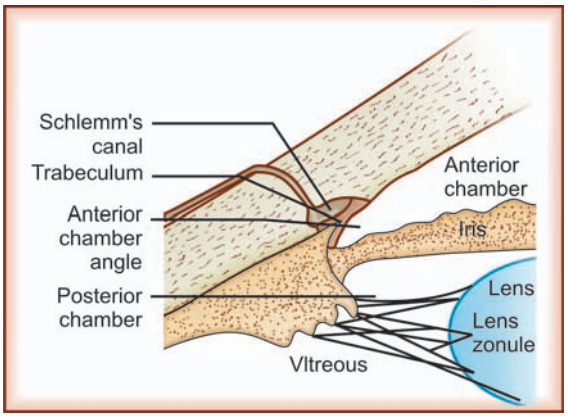


Fig. 15.1: Anatomy of the anterior chamber and its angle

The aqueous humor outflow occurs by two routes trabecular and uveoscleral.

Trabecular Outflow

The bulk of aqueous humor exits the eye through trabecular meshwork-Schlemm's canal-venous

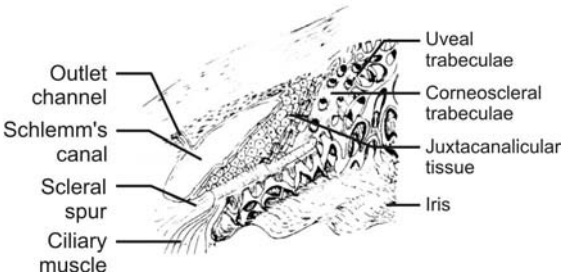


Fig. 15.2: Trabecular meshwork

system route. The trabecular meshwork is composed of multiple layers of connective tissue covered by continuous endothelial layers. Trabeculum is the site for *pressure-dependent aqueous outflow* functioning as a one way valve that allows aqueous to leave the eye but does not allow the flow inside it.

Trabecular meshwork is anatomically divided into 3 parts (Fig. 15.2):

i. *Uveal meshwork:* It is the smaller and innermost part of the trabeculum. It is arranged in bands that extend from the iris root and ciliary body to the Schwalbe's line. The uveal meshwork has larger openings measuring 25 to 75 μ in diameter.
ii. *Corneoscleral meshwork:* It is the larger part of the trabeculum which extends between scleral spur and the lateral wall of the scleral sulcus. It is composed of circumferentially disposed flattened bands with criss-cross arrangements. The corneoscleral meshwork has multiple small openings measuring 5 to 50 μ.
iii. *Juxtacanalicular meshwork:* It is the outermost part of the trabeculum lined on either side by endothelium. It lies adjacent to Schlemm's canal and actually forms the inner wall of the canal. This part of the trabeculum is thought to be the major site of outflow resistance.

Schlemm's Canal

The Schlemm's canal is a single channel with an average diameter of 370 μ, that lies circumferentially in the scleral sulcus. It is lined by endothelium and traversed by tubules. The inner wall of the canal contains giant vacuoles that have direct communication with the intratrabecular spaces. The outer wall of the canal does not contain any pores. A complex system of vessels connects the canal to the episcleral veins. The intrascleral vessels (aqueous veins) may form a direct connection with episcleral veins or may form an intrascleral plexus and then join the episcleral veins.

Uveoscleral Outflow

Besides the conventional trabecular outflow, the aqueous humor exits the eye through uveoscleral route as well. The uveoscleral outflow is also known as *pressure-independent outflow*. It has been estimated to account for 5 to 15% of the total aqueous outflow. The aqueous passes into the ciliary muscles and then into the supraciliary and suprachoroidal spaces.

Visualization of the Angle of the Anterior Chamber

Gonioscopy is performed to visualize the structures as well as the width of the anterior chamber angle. The angle can be described either by using a standard grading system or drawing the iris contour, localization of iris insertion and angle between the iris and the trabecular meshwork. The Shaffer's system of grading the angle (Table 15.1) is commonly used and described below:

All forms of glaucoma are classified into primary and secondary. When no etiology and pathomechanism are found, a *primary glaucoma* is considered. However, when pathomechanisms are evident the disease is categorized as *secondary glaucoma*.

Table 15.1: Shaffer's grading of the angle of the anterior chamber

Grade	*Anterior chamber angle*	*Angle width in degrees*	*Structures visible*	*Chance of closure*
IV	Wide Open (Fig.15.3)	45	Schwalbe's line to ciliary body	Nil
III	Open (Fig.15.4)	20 to <45	Schwalbe's line to scleral spur	Nil
II	Moderately narrow	20	Schwalbe's line to trabeculum	Possible
I	Very narrow (Fig.15.5)	10	Schwalbe's line only	High
0	Closed	0	None	Already closed

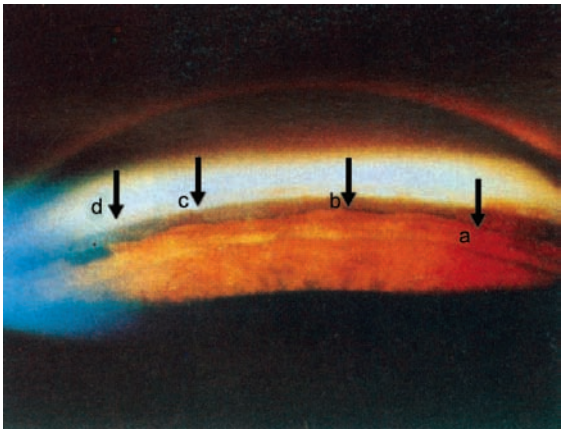


Fig. 15.3: Wide open angle of the anterior chamber (Grade IV) a: Iris root, b: Ciliary body, c: Scleral spur, d: Trabecular meshwork, (*Courtesy*: Drs A Narayanswamy and L Vijaya, Sankara Nethralaya,Chennai)

Fig. 15.4: Open-angle of the anterior chamber (Grade III) (Courtesy: Drs A Narayanswamy and L Vijaya, Sankara Nethralaya, Chennai)

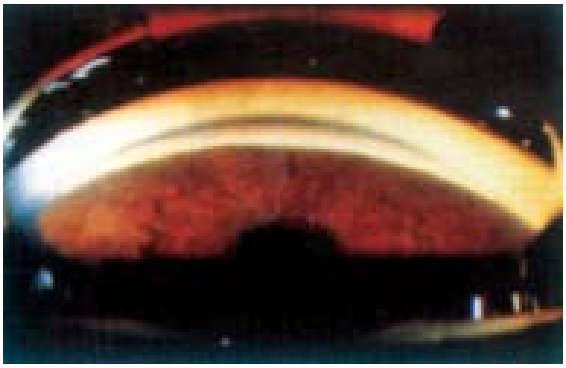


Fig. 15.5: Very narrow-angle of the anterior chamber (Grade I) (*Courtesy*: Drs A Narayanswamy and L Vijaya, Sankara Nethralaya, Chennai)

Classification of Glaucomas

1. *Developmental glaucomas*
   a. Congenital glaucoma (Buphthalmos)
   b. Infantile glaucoma
   c. Juvenile glaucoma
   d. Developmental glaucoma associated with congenital anomalies
2. *Primary open-angle glaucoma (POAG)*
   a. Primary open-angle glaucoma with high pressure
   b. Primary open-angle glaucoma with normal pressure
3. *Primary angle-closure glaucoma (PACG)*
4. *Secondary glaucomas.*

Terminology

Developmental glaucoma: The term developmental glaucoma includes primary congenital glaucoma and glaucoma associated with ocular or systemic developmental anomalies.

Congenital glaucoma: Glaucoma that manifests at birth or during the first year of life.

Infantile glaucoma: When glaucoma occurs within first few years of life.

Juvenile or *childhood glaucoma:* When glaucoma occurs between 3 and 16 years of age, it is labeled as juvenile or childhood glaucoma.

Secondary infantile glaucoma: When the rise of IOP is associated with inflammatory and neoplastic conditions of the eye or metabolic disorders, it is called *secondary infantile glaucoma*.

PRIMARY CONGENITAL GLAUCOMAS

Congenital Glaucoma (Buphthalmos)

Congenital glaucoma or buphthalmos is an early onset primary congenital glaucoma characterized by dysgenesis of the angle of the anterior chamber, raised IOP, corneal opacities and enlargement of the globe (Fig. 15.6). When the IOP is elevated before the age of 3 years, it results in enlargement

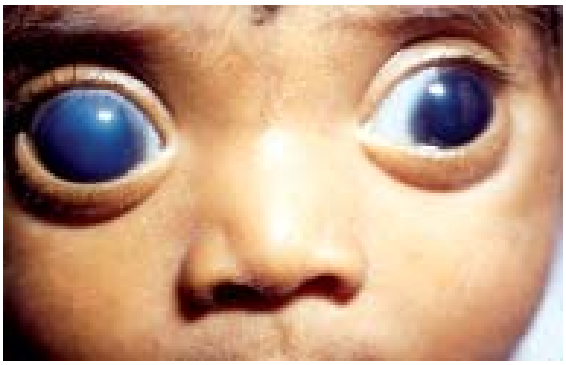


Fig.15.6: Bilateral buphthalmos
(*Courtesy:* Dr AK Mandal, LVPEI, Hyderabad)

of the globe. The enlarged eye due to congenital glaucoma is often referred to as *buphthalmos*.

Etiology

Primary congenital glaucoma is usually sporadic, only 10 percent of cases show an autosomal recessive inheritance with variable penetration. Chromosomal abnormalities have been reported at locations 1p36 and 2q21. The disease occurs more frequently in male infants (65%) than female. It is caused by maldevelopment of the angle of the anterior chamber (angle dysgenesis). The presence of a continuous cellular membrane in the angle of the anterior chamber may also obstruct the drainage of aqueous humor.

The characteristic gonioscopic appearance of an eye with congenital glaucoma is marked by the presence of open angle, Barkan's membrane (trabeculodysgenesis), abnormally high insertion of the iris, poorly developed and posteriorly placed scleral spur and a collapsed Schlemm's canal.

Clinical Features

Photophobia, lacrimation and blepharospasm form the classical triad of symptoms of buphthalmos. The child is often irritable. The buphthalmic eye usually presents a mild degree of proptosis with enlargement of globe owing to extensibility of the sclera in infants. The ocular tension is markedly raised and produces edema of the cornea. At a later stage, discrete stromal opacities appear as double contour lines (Haab's striae) due to the rupture of Descemet's membrane. The sustained elevation of intraocular pressure causes further stretching of globe and the cornea becomes enlarged.

Buphthalmos must be differentiated from keratoglobus and megalocornea. Measurement of intraocular pressure, gonioscopy and evaluation of optic nerve head are helpful in making the diagnosis (Table 15.2). The anterior chamber is usually deep and the angle anomalies are obvious on gonioscopy.

Table 15.2: Differentiating features of buphthalmos, keratoglobus and megalocornea

Features	*Buphthalmos*	*Keratoglobus*	*Megalocornea*
Laterality	Usually bilateral	Bilateral	Almost always bilateral
Sex predilection	M:F :: 5:3	Uncertain	Male (90%)
Hereditary pattern	Autosomal recessive	Uncertain	X-linked recessive
Vision	Markedly impaired	Impaired	Not impaired
Corneal transparency	Opacities due to rupture in Descemet's membrane	Stromal haze due to fragmentation of Bowman's membrane	Transparent
Intraocular pressure	Raised	Normal	Normal
Angle of anterior chamber	Developmental anomalies seen	Normal	Normal
Optic disk	May be cupped	No cupping	No cupping

The later stages of the disease may show congestion in the conjunctiva, apparent bluish discoloration of sclera at the ciliary region owing to shining of the underlying uveal pigments, iris atrophy, tremulous iris, luxation of the lens and marked cupping of the optic nerve head. As a result of the increased axial length, the eye usually becomes myopic. Nevertheless, it may be compensated due to flattening of the cornea, and flattening and backward displacement of the lens.

A definite diagnosis of congenital glaucoma requires meticulous examination of the infant under general anesthesia. It includes recording of the intraocular pressure by hand-held applanation tonometer, gonioscopic assessment of the angle of the anterior chamber of the two eyes by Koeppe's lens or Richardson modification of Koeppe's lens, examination of the optic nerve head for cupping and measurement of the corneal diameter. A corneal diameter greater than 12 mm before the age of one year is highly suspicious of congenital glaucoma.

The clinical features of infantile glaucoma, which manifests during the first few years of life, are almost same as that of congenital glaucoma, but it occurs later because the angle of the anterior chamber is more mature than when glaucoma is present at birth.

Treatment

Anti-glaucoma medications have limited value in the management of congenital and infantile glaucomas and most effective therapy is surgical. However, beta-blockers and carbonic anhydrase inhibitors are often used to reduce IOP in the preoperative period.

Goniotomy or trabeculotomy is the initial procedure of choice in patients with clear cornea.

Trabeculotomy *ab externo* is preferred in patients with opaque cornea. When initial procedure fails, trabeculectomy with or without mitomycin C (MMC) should be performed.

Prognosis

Long-term results of early surgical intervention of congenital glaucoma have greatly improved, however, late complications such as corneal scarring, amblyopia, and cataract are common.

Juvenile or Childhood Glaucoma (Late Onset Primary Congenital Glaucoma)

Sometimes the primary congenital glaucoma may manifest between the third and the sixteenth year of life. The basic defect lies in the angle of the anterior chamber. The structures are poorly differentiated, hence decreased aqueous outflow occurs. The child remains symptom-free and often presents late with visual loss especially the visual field defects. The IOP remains moderately elevated but the eye never shows enlargement and corneal changes. The evaluation of optic nerve head may reveal enlargement of optic cup and diffuse thinning of the neuroretinal rim.

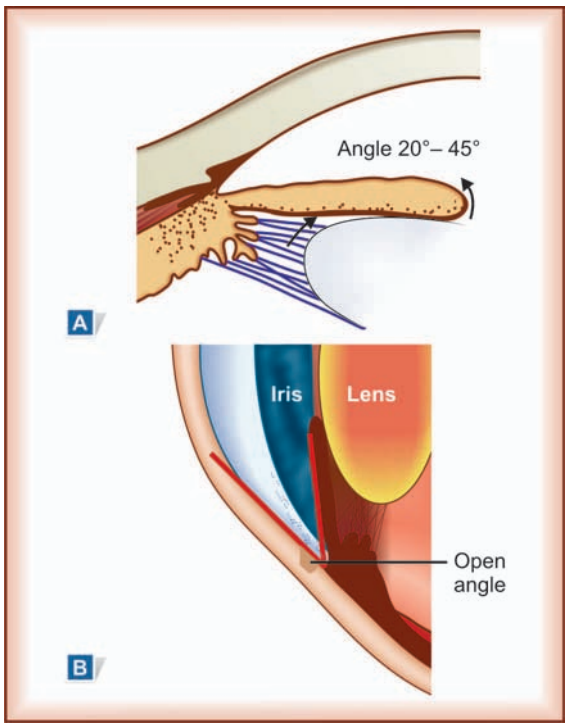
The congenital glaucoma of late onset needs surgical management. Trabeculotomy or trabeculectomy provides satisfactory results.

Developmental Glaucoma Associated with Congenital Anomalies

Developmental glaucomas may be associated with Peter's anomaly, Rieger's anomaly, Axenfeld anomaly, microcornea, aniridia, ectopia lentis, and nanophthalmos. It may also manifest as a part of a number of ophthalmic syndromes such as Sturge-Weber syndrome, Marfan's syndrome, rubella syndrome and Lowe's syndrome.

PRIMARY OPEN-ANGLE GLAUCOMA

Primary open-angle glaucoma (POAG) is defined as a chronic progressive optic neuropathy associated with elevated IOP and visual field defects. It is relatively more common than primary angle-closure glaucoma and affects nearly 1% of the population over the age of 40 years.



Figs 15.7A and B: Open angle of the anterior chamber

Etiology

The etiology of POAG is obscure. The disease occurs without any precipitating or pre-existing ocular or systemic disease. As the name denotes, it occurs in eyes with open-angle of the anterior chamber (Figs 15.7A and B). The intraocular pressure increases due to a decrease in the aqueous outflow across the trabecular meshwork owing to trabecular sclerosis and loss of cells.

Risk Factors of POAG

Risk factors of POAG include IOP, central corneal thickness (CCT), age, race, family history of glaucoma, myopia, central retinal vein occlusion (CRVO), cardiovascular diseases, diabetes, and hypertension.

IOP: An average mean normal normal IOP of 16 ± 3 mm Hg is reported on population-based studies. IOP above 21 mm Hg is often considered as abnormal.

CCT: It affects the measurement of IOP. An average central corneal thickness is 544 µ in normal eyes. It has been estimated that IOP increases at 2-7 mm Hg per 100 µ increase in the corneal thickness. A decrease in corneal thickness is observed in patients with normal pressure glaucoma.

Age and *Race*: Age is another important risk factor for glaucoma. The prevalence of glaucoma increases with advancement of age. The disease is more prevalent in certain races like black Africans.

Heredity and *Family history:* The disease is usually inherited in a multifactorial manner with variable penetrance. It is familial and nearly 10% of the first degree relatives (siblings and offsprings) of patients with glaucoma eventually develop the disease.

Myopia: It has been reported that individuals with myopia may be at a greater risk for the development of POAG.

CRVO: Patients with CRVO may present with concomitant POAG or vice-versa.

The rise of intraocular pressure in primary open-angle glaucoma is probably caused by interference with aqueous outflow owing to degenerative changes in the trabeculum, Schlemm's canal and exit channels. The optic nerve damage (cupping) may also be caused by accompanying vascular insufficiency. Such a concept is supported by an observation that the cupping sometimes continues to progress even after the normalization of intraocular pressure by medical therapy or surgery.

Clinical Features

Generally, POAG is a bilateral symptom-free chronic condition having a slow progressive course. Mild headache, eye or browache, difficulties in reading or doing close work and frequent

changes of presbyopic glasses are the usual presenting symptoms of the disease.

Occasionally, the disease is so insidious that it is not noticed until the vision of the affected eye is seriously impaired and extensive visual field defects ensue. The slow and silent course of the disease has earned the name *chronic simple glaucoma*. Since the disease usually starts after the age of 40 years, when the presbyopic symptoms start, a detailed check-up to exclude glaucoma is needed whenever reading glasses are being prescribed.

Open-angle glaucoma affects the emmetropic or myopic eye having normal depth of the anterior chamber. Initially, the pupil is briskly reacting to light but later becomes sluggish. The diagnosis of the disease depends on raised IOP, cupping of the disk, and visual field defects.

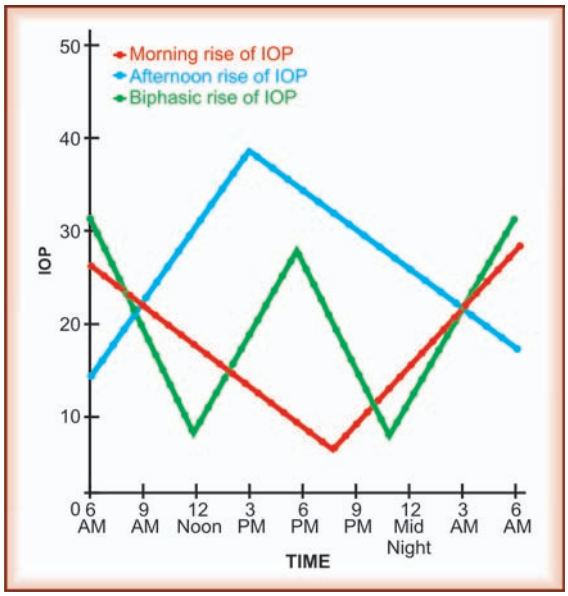


Fig.15.8: Diurnal variations in IOP

Intraocular Pressure

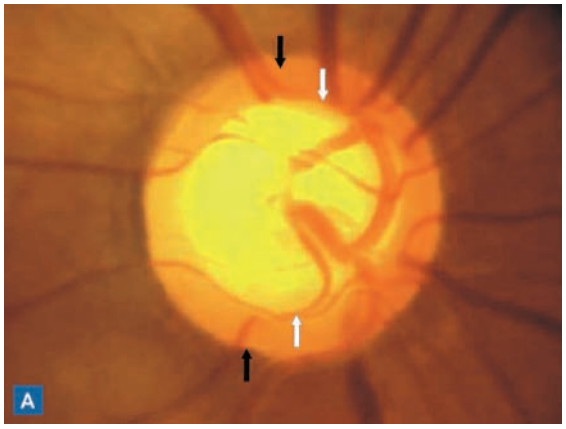
An applanation pressure of 21 mm Hg has been arbitrarily fixed as the dividing line between normal and abnormal tension. The ocular pressure shows great variations in open-angle glaucoma and, therefore, requires careful tonometry.

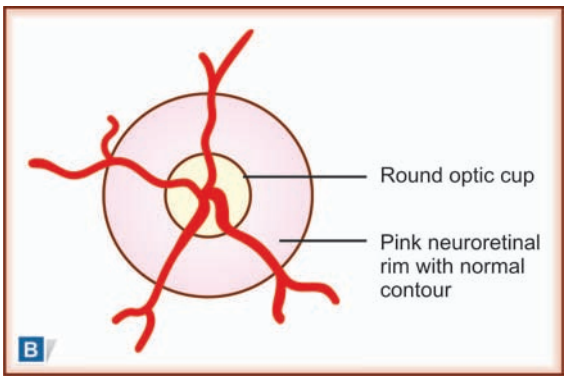
Diurnal variations in IOP: Initially, the IOP may not show a rise but only an exaggeration of the normal diurnal variation. The diurnal variation of intraocular pressure in normal eye has a mean of 3.7 mm Hg, and majority of the subjects have a variation of IOP between 2 mm Hg and 6 mm Hg over a 24 hours period, due to aqueous humor production changes. The IOP shows variations during different periods of a day. It often follows a circadian cycle with rise in the morning (8 to 11 AM) and fall in the night (12 midnight to 2 AM). The fluctuation in IOP is sleep-cycle dependant rather than daylight-cycle dependant. The IOP may show a morning rise, or afternoon rise, or a biphasic rise (Fig.15.8) in some patients with POAG. Higher IOP is associated with greater fluctuations. A diurnal fluctuation of greater than 10 mm Hg is suggestive of glaucoma.

The disease may present with asymmetrical levels of intraocular pressure. Although it is a bilateral disease, the condition is often more advanced in one eye. In the early phase of the disease the tension usually returns to normal between the phasic rises, but after sometimes the normal level is no longer attained and the tension causes visual loss, damage to the optic nerve fibers and consequent visual field defect. The measurement of intraocular pressure alone is insufficient to diagnose open-angle glaucoma.

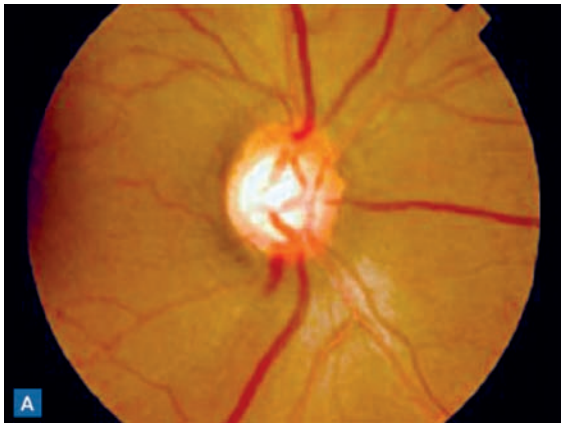
Optic Nerve Head and Retinal Nerve Fiber Layer

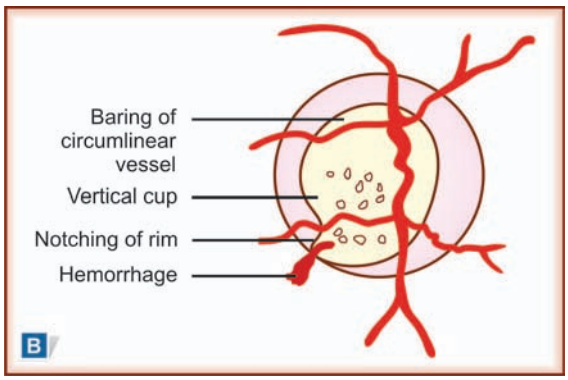
The evaluation of the optic nerve head (ONH) and retinal nerve fiber layer (RNFL) is essential in the diagnosis of POAG. Earlier, pathological cupping of the optic nerve head was considered the most significant diagnostic sign of the disease. As the size of the cup varies physiologically with the overall size of the disk, the cupping of the disk has limited value. Normally, the physiological cup occupies less than 30% area of the optic disk (Figs 15.9A and B) and is symmetrical.





Figs 15.9A and B: (A) Normal optic nerve head (*Courtesy*: Dr Chandra Sekhar LVPEI, Hyderabad), (B) Diagrammatic representation of normal optic cup and neuroretinal rim



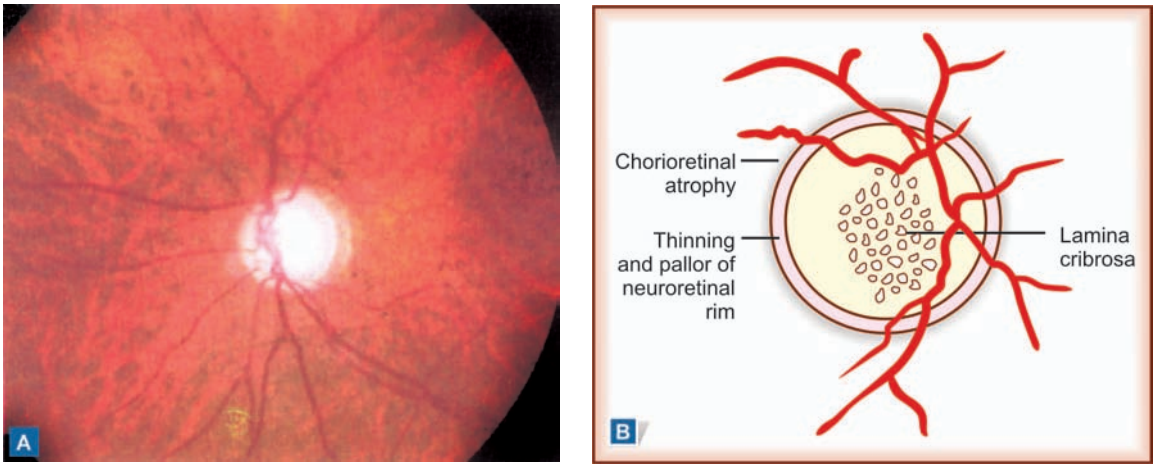


Figs 15.10A and B: (A) Glaucomatous cup, (B) Diagrammatic representation of vertically enlarged cup with notching of neuroretinal rim and splinter hemorrhage

The vertical cup/disk ratio is a relatively better measure of deviation from normal than the horizontal ratio because neuroretinal rim loss occurs early at the upper and lower poles of the disk. A cup/disk ratio of more than 0.65 is found in less than 5% of the normal population. Asymmetry of cup/disk ratio greater than 0.2 between the two eyes is generally seen in nearly 70% of patients with POAG. The progressive loss of the nerve fibers nasally causes a nasal displacement of the retinal blood vessels. The extension of cup posteriorly results in double angulation of the blood vessels *(bayonetting sign)* in which the vessels dip sharply backwards then pass over the steep walls of the cup before angling again on the floor of the cup.

An advanced glaucomatous cupping is characterized by depression of the lamina cribrosa, vertical oval shape, thinning or absence of the neuroretinal rim, displacement of central retinal vessels and pallor of the disk (Figs 15.10A and B and 15.11A and B). The cup of the optic disk enlarges in extent and depth, due to the disappearance of the optic nerve fibers without any glial proliferation, and results in the formation of large caverns (cavernous optic atrophy).

Neuroretinal Rim: The tissue between the cup and disk margin is known as *neuroretinal rim (NRR).* Normally it has an orange or pink color. The rim is widest in the inferior disk region followed by



Figs 15.11A and B: (A) Pale atrophic optik disk with advanced glaucomatous cup, (B) Diagrammatic representation of advanced glaucomatous cupping with very narrow neuroretinal rim

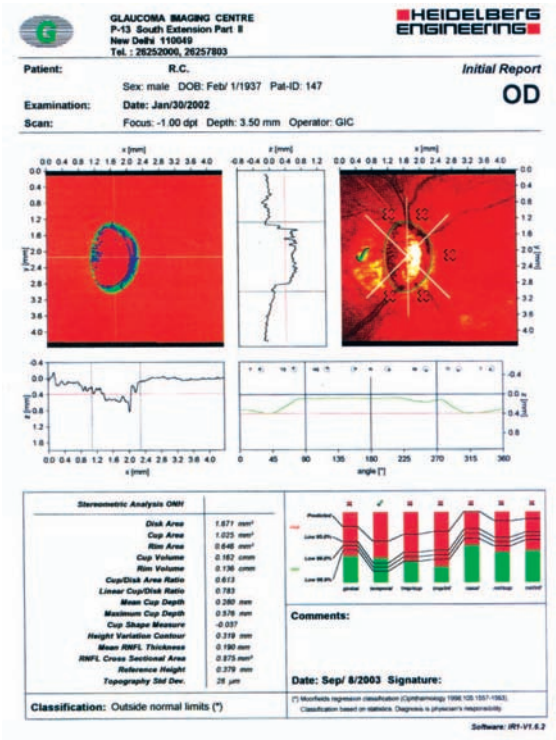


Fig.15.12: Typical baseline HRT tomography scan analysis of optic nerve head (*Courtesy*: Dr D Sood, Glaucoma Imaging Centre, New Delhi)

the superior, the nasal and the temporal disk region (ISNT rule, Fig. 15.9A).

Focal enlargement of the cup causes notching or narrowing of the rim typically seen at the inferior or superior temporal poles of the disk in the early glaucomatous optic neuropathy. Notching of the rim at the upper and lower poles of the disk is responsible for vertical oval shape of

the cup (Fig. 15.10B). Heidelberg retina tomogram (HRT II) can be used to obtain stereometric analysis of the optic neve head. It measures the area and volume of the optic disk, cup, rim (Fig. 15.12) and mean thickness of the retinal nerve fiber layer.

Splinter hemorrhage: Besides cupping and notching or thinning of neuroretinal rim, splinter hemorrhage (Fig. 15.10B) may be seen on or near the disk in approximately one third of the glaucomatous patients. Patients with normal tension glaucoma are more prone to have disk hemorrhage. These patients with splinter hemorrhage are more likely to develop a progressive visual field loss. Such hemorrhage may also be found in patients with posterior vitreous detachment,diabetes and branch retinal vein occlusion.

Peripapillary Atrophy (PPA): Peripapillary atrophy (Fig. 15.13) is found in a greater frequency and is more extensive in eyes with glaucoma than in normal eyes. The localized PPA results in corresponding visual field defects.

Nerve Fiber Layer Defects: The nerve fiber layer has a refractile appearance with fine striations created by bundles of axons. It shows focal or diffuse abnormalities (Fig. 15.14) in glaucomatous neuropathy. Focal abnormalities include slit-grooves or slit-defects. The diffuse nerve fiber loss is more common in glaucoma than the focal loss. The early nerve fiber loss occurs as translucency of the neuroretinal rim visible on slit-lamp biomicroscopy. The quantitative loss of nerve fiber layer can be made out by using GDx nerve fiber analyzer (Fig. 15.15) or optical coherence tomography.

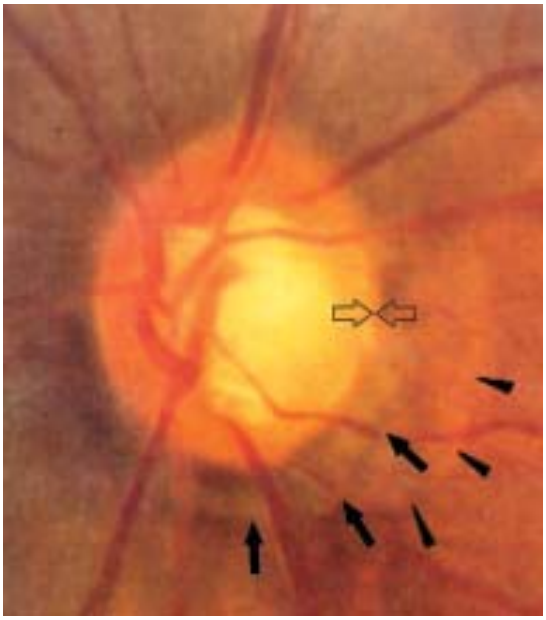


Fig.15.13: Peripapillary atrophy
(*Courtesy*: Dr Ki Ho Park, National University, Seoul)

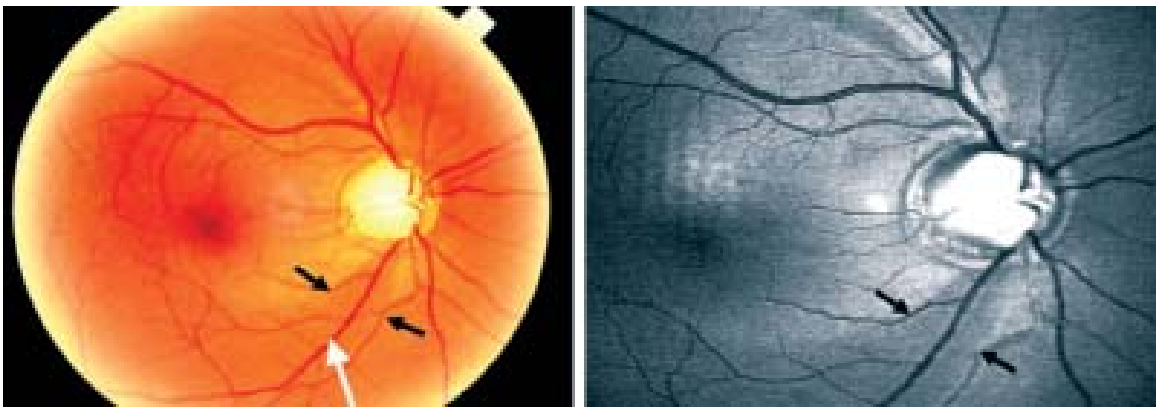


Fig.15.14: Focal nerve fiber layer defect (*Courtesy*: Dr R Parikh and G Chandra Sekhar, LVPEI, Hyderabad)

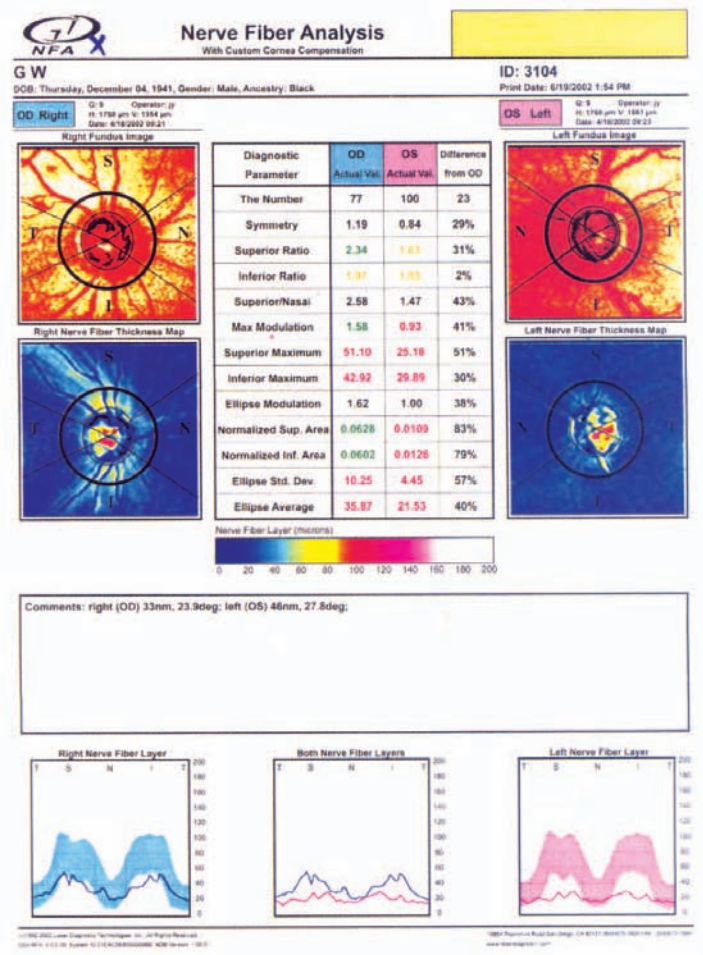


Fig.15.15: Retinal nerve fiber data obtained from a GDx nerve fiber analyzer (*Courtesy:* Dr D Sood, Glaucoma Imaging Centre, New Delhi)

Causes of Optic Nerve Damage in Glaucoma

Besides elevated IOP, the development of glaucomatous optic neuropathy results from a number of factors. Two hypotheses—mechanical and ischemic—have been propogated.

The *mechanical theory* advocates direct compression of axonal fibers and supporting structures of the optic nerve resulting in bowing backward of lamina cribrosa and interruption in the axoplasmic flow and death of retinal ganglion cells .

The *ischemic theory* lays stress on the development of intraneural ischemia due to decrease in optic nerve perfusion.

Perhaps, both mechanical and ischemic factors are responsible for the optic nerve damage .

Ideally stereoscopic colored photographs are useful for documentation of optic neuropathy. If fundus camera is not available, drawing must be made. Every glaucomatous cupping must be differentiated from physiological cupping and the cupping of primary optic atrophy. The physiological cup is funnel-shaped with sloping edges.

It is localized, and the disk retains its pink color. The cup of primary optic atrophy is saucer-shaped and shallow, and the disk appears white. The arteriosclerotic optic atrophy may present typical cupping of the disk and the same visual field defects as those in open-angle glaucoma.

Visual Field Defects

The visual field defects in open-angle glaucoma are due to the damage of optic nerve fiber bundles and run more or less parallel to the degenerative changes in the optic nerve fibers.

The nerve fibers from retina pass to the optic nerve head in a set pattern. The fibers nasal to optic disk take a straight course to reach the nasal side of the disk. The fibers from macula (papillomacular bundle) run directly to the temporal margin of the disk. The macular fibers are resistant to glaucomatous damage. Therefore, central island of visual field is retained even in advanced glaucoma. The fibers from the retina temporal to macula extend in an arcuate manner around the papillomacular bundle to reach the upper and lower poles of the optic disk (Fig. 15.16).The arcuate nerve fibers are most sensitive to glaucomatous damage resulting in early arcuate field defects. In glaucoma, usually the lower nerve fibers are affected earlier than the upper fibers.

Visual fields are recorded with the help of either manual or automated perimeter. The latter is preferred as it is more reliable, repeatable and provides statistical analysis. It measures the retinal sensitivity or threshold at different locations by varying the brightness of test target. Humphrey perimeter programs 24-2 or 30-2 threshold test (Fig. 15.17) and Octopus program G1 (Fig. 15.18) are considered standard tests for glaucoma.

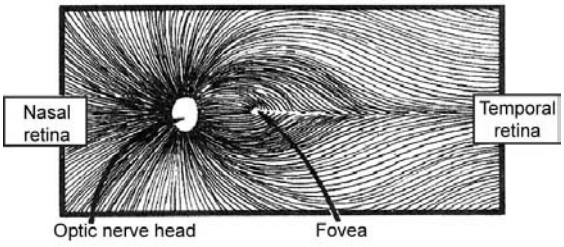


Fig. 15.16: Arrangement of retinal nerve fibers

The visual field defects in POAG include (i) generalized depression, (ii) paracentral scotoma, (iii) arcuate scotoma, (iv) nasal step, (v) altitudinal defect and (vi) temporal wedge.

Both central and peripheral fields should be determined under standardized conditions.

A generalized depression in the visual field may occur with diffuse glaucomatous damage. It can also occur with opacities in the ocular media.

The glaucomatous field defects manifest in stages. The most common early visual field defects in glaucoma are small isolated *paracentral scotomas* between 2 and 10 degrees (Figs 15.19A and B). Initially, these scotomas are relative, but eventually they become absolute. The visual field defects must be clinically correlated with changes in the optic nerve head.

In uncontrolled glaucoma, a *sickle-shaped extension of the blind spot* (Fig. 15.20), above or below, with the concavity towards the fixation point may develop, known as *Seidel's sign*.

Isolated scotomas in central field (Bjerrum's area) may coalesce and form a classic arcuate scotoma (Figs 15.21A and B). The *arcuate scotoma* may develop either above or below the horizontal raphe. When there are arcuate scotomas both above and below the horizontal meridian, they join to form a *ring scotoma* or *double arcuate scotoma* (Figs 15.22A and B). Arcuate or Bjerrum's scotomas are usually associated with glaucoma. They can also be found in other conditions such as sudden drop of blood pressure, coronary thrombosis, opticochiasmatic arachnoiditis, pituitary adenoma and drusen of the optic nerve head.

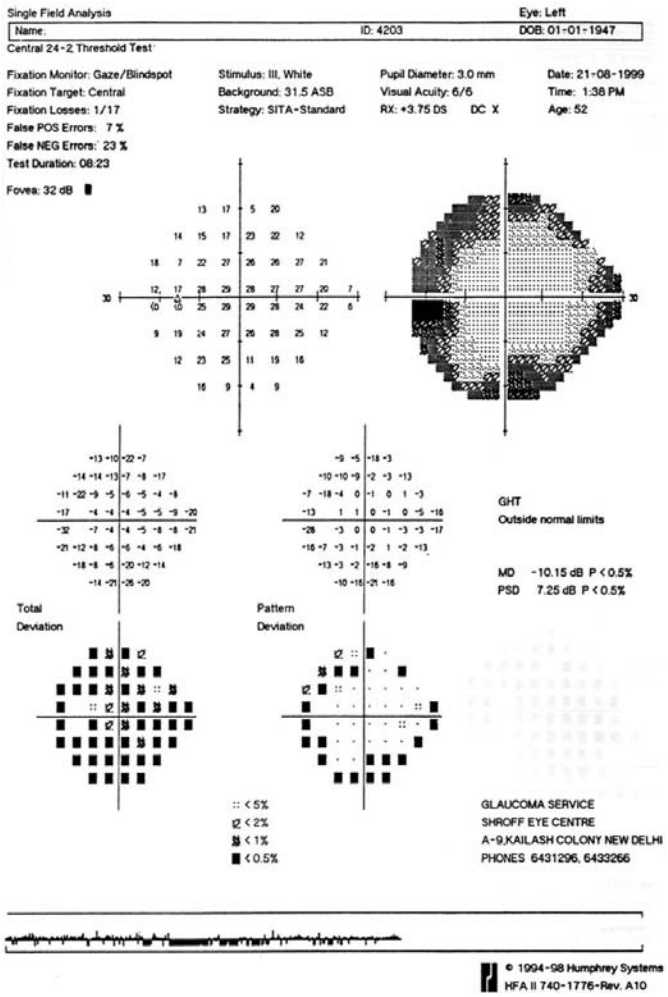


Fig. 15.17: Glaucomatous visual field defects on Humphrey perimeter (*Courtesy*: Dr D Sood, Glaucoma Imaging Centre, New Delhi)

Unequal contraction of peripheral isopters due to the loss of corresponding bundles of peripheral arcuate nerve fibers causes *Roenne's nasal step* (Figs 15.23A and B). In fact, the nasal step delineates the nasal border of the completed arcuate scotoma wherein a sectorial defect in the upper or lower peripheral field presents a sharply defined edge.

The shape of the nasal step varies with the proximity to the fixation point. Its presence confirms the glaucomatous field loss. The altitudinal defects with more or less complete loss of the superior visual field characterize advanced glaucomatous optic neuropathy. The visual field loss in uncontrolled glaucoma gradually spreads both centrally as well as peripherally. Ultimately, only a small island of central vision and an accompanying temporal island are left.

Enlargement of the blind spot, baring of the blind spot and generalized constriction of the

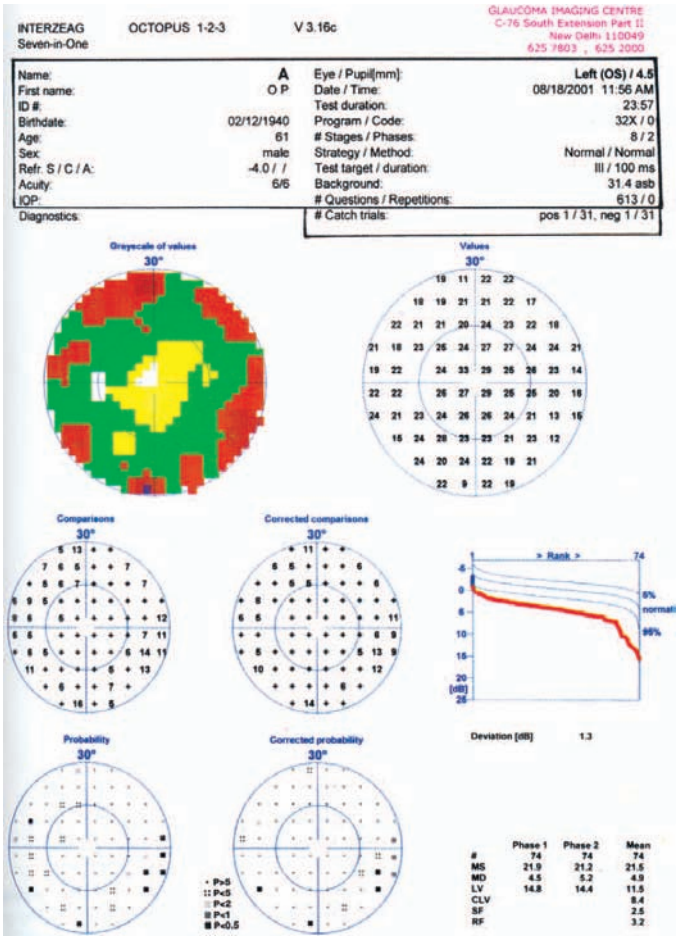


Fig. 15.18: Glaucomatous visual field defects on Octopus perimeter (*Courtesy*: Dr D Sood, Glaucoma Imaging Centre, New Delhi)

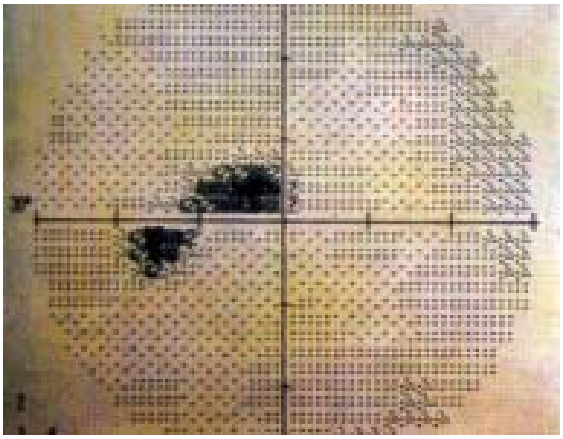


Fig.15.19A: Superior paracentral scotoma correlates well with optic nerve head changes in Fig. 15.19B

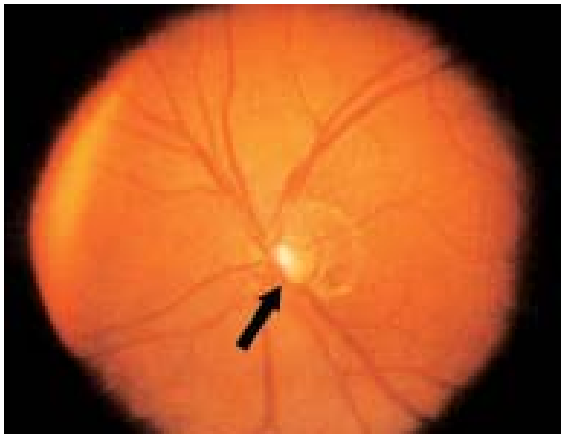


Fig.15.19B: Small optic disk with inferior notch (*Courtesy:* Dr G Chandra Sekhar, LVPEI, Hyderabad)

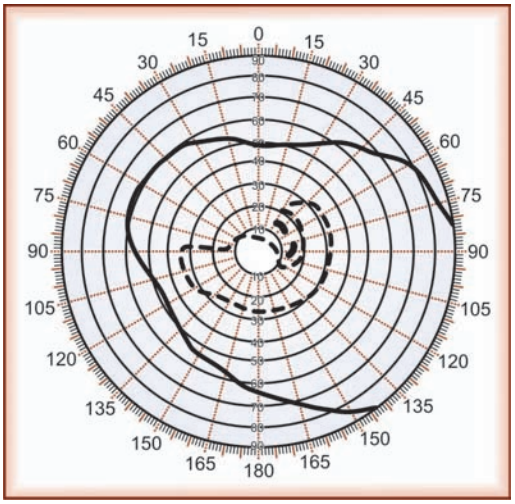


Fig.15.20: Seidel's sign

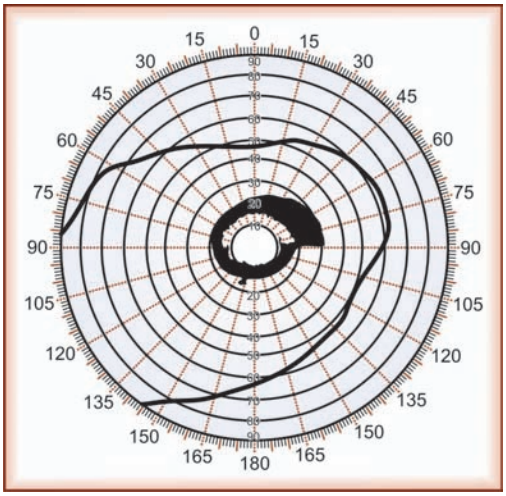


Fig.15.22A: Arcuate scotoma

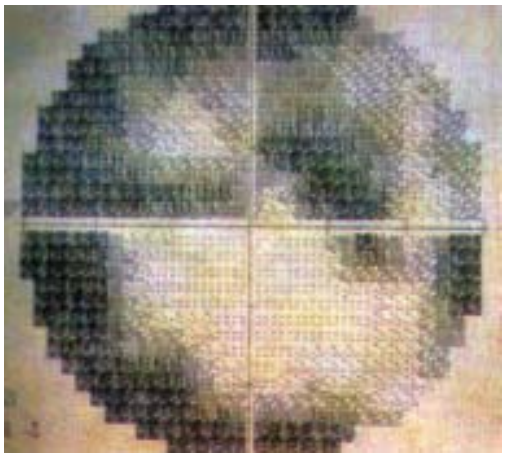


Fig. 15.21A: Superior arcuate scotoma

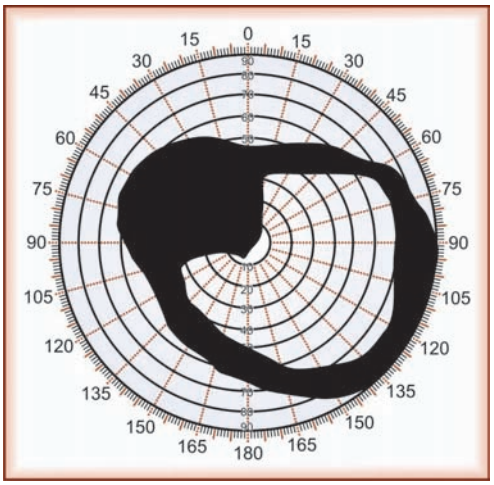


Fig.15.22B: Double arcuate scotoma and quadrantic defect

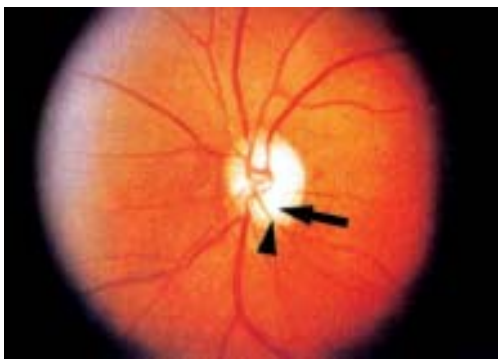


Fig. 15.21B: Optic cup is enlarged and inferior neuroretinal rim is pale and narrow (*Courtesy:* Dr G Chandra Sekhar, LVPEI, Hyderabad)

visual field were formerly considered as early glaucomatous defects. These field defects may be found in other ocular conditions as well. They are no more considered as diagnostic of glaucoma. When visual field is charted manually with a very small test object (1/2000), the central field may show a localized constriction to exclude the blind spot (baring of the blind spot). The baring of the blind spot is a defect not specific enough to be relied on for early detection of glaucoma. It may be found in ageing, miosis, and lens opacities.

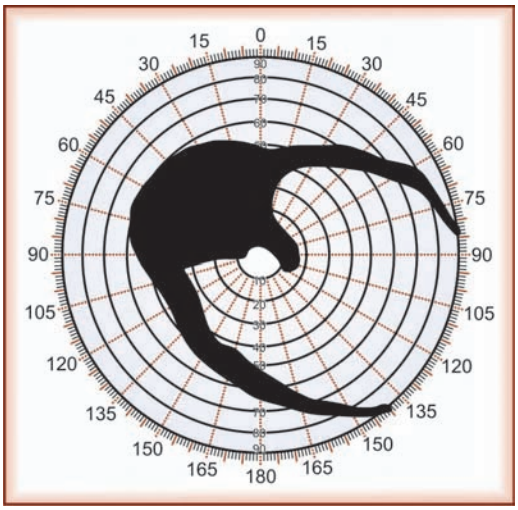


Fig. 15.23A: Roenne's step

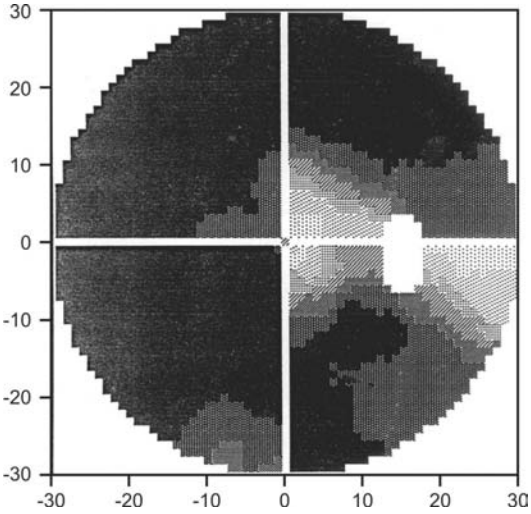


Fig. 15.23B: Both upper and lower arcuate scotomas merging with nasal step

Visual field loss is considered significant when following field defects are present on Humphrey perimetry:

1. Glaucoma Hemifield Test (5 groups of test points in hemifields) is found abnormal on two consecutive tests.
2. A cluster of three points not contiguous with blind spot decline ≥ 10 dB, and
3. Corrected pattern standard deviation remains less than 5% on two consecutive examinations.

Periodic visual field testing (Fig. 15.24) is recommended for patients with glaucoma to assess deterioration in the visual field over a period of time with a view to re-evaluate the desired target pressure and modify treatment.

Normal Tension Glaucoma

Whether normal tension glaucoma (NTG) is a separate disease entity or a type of POAG remains an unresolved controversy.

Etiology

In NTG, IOP remains normal but other risk factors such as ischemic vascular diseases, vasospastic migraine, autoimmune diseases and coagulopathies play more important role.

Clinical Features

NTG presents characteristic signs of POAG despite low or normal IOP. The disease is progressive and causes optic nerve damage. The neuroretinal rim is thinner especially inferiorly and inferotemporally. Splinter optic disk hemorrhages (40%) are frequent. Varied patterns of peripapillary atrophy may be found. The visual field defects in NTG tend to be more focal, deeper and closer to fixation point as compared to that in POAG.

In spite of the name, normal pressure glaucoma, great care must be taken to record the IOP. The low tonometric readings may be due to low scleral rigidity and reduced corneal thickness. Some patients with NTG present asymmetric IOP. It is observed that the eye with higher IOP suffers worst damage.

Differential Diagnosis

Many clinical entities mimic the optic neuropathy and visual field changes found in NTG. These include POAG, coloboma of the optic nerve head, tumor of optic chiasma, anterior ischemic optic neuropathy, optic disk drusen, and shock optic neuropathy.

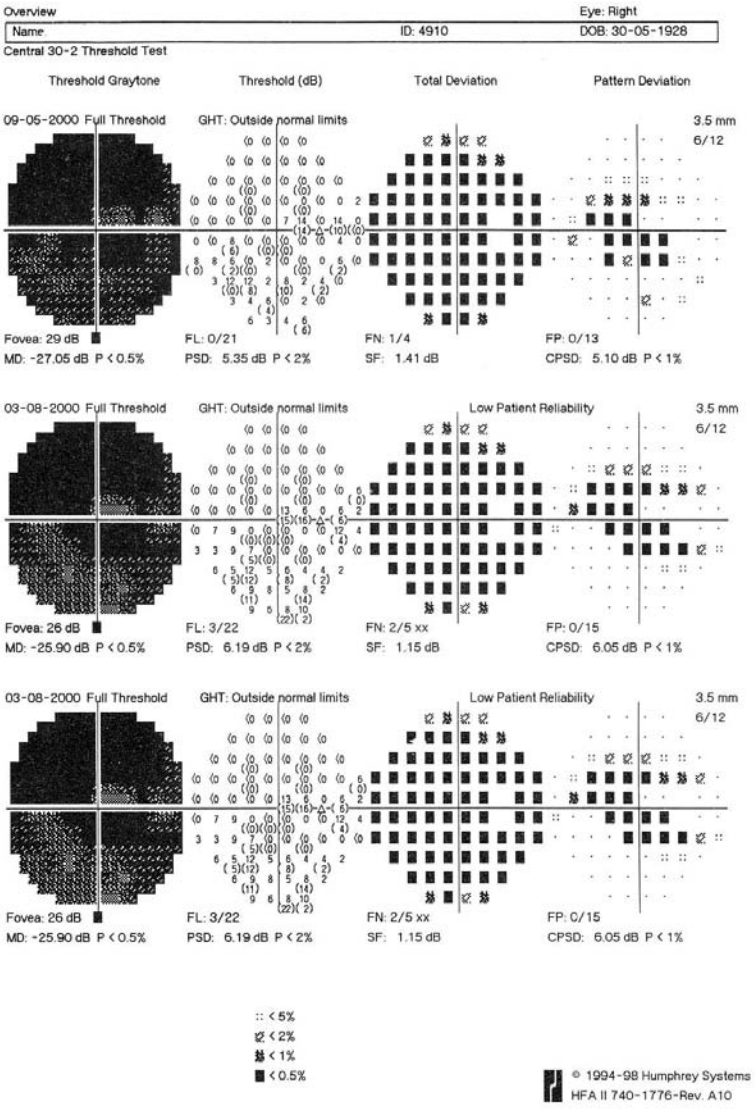


Fig.15.24: Periodic analysis of the visual fields

Treatment

Normal pressure glaucoma is treated on the lines of primary open-angle glaucoma but the target pressure is usually kept relatively low (10-12 mm Hg).

Prognosis

The clinical course of NTG is variable. The disease may not progress in some patients despite the lack of treatment whereas others show deterioration in spite of aggressive reduction in IOP.

Ocular Hypertension

Ocular hypertension or glaucoma suspect is defined as one who has an elevated IOP in the absence of identifiable optic neuropathy and visual field defects.

Ocular hypertension is considered as a benign rise of intraocular pressure usually found in about 6 to 10% of population above 40 years of age and is more common than open-angle glaucoma (0.3-0.5% of the same population). However, long-term follow-up studies (5 years) have shown that nearly 5% of cases of ocular hypertension may develop glaucoma.

Ocular Hypertension Treatment Study revealed that in spite of reduction of IOP by topical anti-glaucoma medication, 4.4% of patients with ocular hypertension progress to glaucoma as compared to 9.5% of patient in untreated group. Higher base-line IOP, reduced central corneal thickness, and increased cup-disk ratio are important risk factors for the development of glaucoma in patients with ocular hypertension.

Therefore, some ophthalmologists advocate to drop the term ocular hypertension from the literature and prefer to use the term *glaucoma suspect* in order to stress the need for long-term follow-up.

The intraocular pressure in ocular hypertension varies widely between 20 and 40 mm Hg. The patient with tension between 20 and 25 mm Hg does not need any treatment unless associated with risk factors like POAG in the fellow eye, diabetes, thyroid dysfunction, asymmetry of optic cup and family history of glaucoma. However, a careful follow-up is necessary at 6-monthly interval. Patients with an intraocular pressure of more than 30 mm Hg need medical management.

Diagnosis of POAG

Glaucoma can produce serious visual impairment through destruction of the optic nerve fibers. It is, therefore, necessary that the disease should be diagnosed before it causes irreversible visual damage. The early diagnosis of glaucoma requires an awareness on the part of ophthalmologists coupled with routine screening of all subjects over the age of 40 years. Following tests help in the early diagnosis of POAG.

1. *IOP*: Measurements of intraocular pressure preferably by applanation tonometer.
2. *Diurnal variation in IOP:* The IOP should be measured several times in a day to detect fluctuations. In normal individuals, IOP fluctuations vary between 2 and 6 mm Hg over a 24-hour period. Diurnal variation of greater than 10 mm Hg is suggestive of glaucoma.
3. *Central Corneal Thickness*: Corneal pachymetry should be carried out to measure the central corneal thickness. CCT affects the IOP measurement. An increase in CCT gives an erroneously high IOP reading. A decrease in CCT gives low IOP measurements.
4. *Slit-lamp examination* of the anterior segment of the eye is useful in excluding the secondary open-angle glaucoma.
5. *Gonioscopic examination* is essential to differentiate between primary and secondary glaucoma as well as between POAG and PACG.
6. *Evaluation of optic disk*: Careful stereoscopic optic disk evaluation is important to rule out physiological enlarged cup and other congenital and acquired disk anomalies.
7. *Quantitative measurement of the retinal nerve fiber layer*: An objective measurement of RNFL can be performed with the help of confocal scanning laser ophthalmoscope or optical coherence tomography.
8. *Visual fields*: Perimetry or clinical assessment of the visual fileds enables detection of early glaucoma. It is important to correlate changes in the visual field with changes in the optic nerve head.
9. *Provocative test*: In suspicious cases with borderline intraocular pressure, provocative

tests are carried out to establish a degree of probability that a patient does or does not have glaucoma. *Water-drinking test* may be used for this purpose. The patient comes empty stomach and his initial intraocular pressure is recorded. Then he drinks approximately one liter of cool water within a span of five minutes. The intraocular pressure is measured at 15 minutes intervals for one hour or until the pressure stops rising. A rise greater than 8 mm Hg is suggestive of a pathological response seen in patients of open-angle glaucoma.

The rise in intraocular pressure induced by water intake is probably due to the transfer of water from diluted blood into the more concentrated aqueous humor. It is suggested that a positive response of water-drinking test is a function of baseline intraocular pressure and may not be related with presence or absence of glaucoma. The water-drinking test may be combined with tonography. However, the test fails to provide a definitive diagnosis.

Treatment

The main aim of management of open-angle glaucoma is to reduce the intraocular pressure to a level at which it does not produce further damage to the optic nerve fibers. Such a pressure is called as *target pressure*. It is judged by the stabilization of the visual field defects and evaluation of the appearance of optic nerve head. An initial reduction of 20% IOP from the baseline pressure is suggested. However, reduction of IOP to target pressure may not ensure that progression in glaucomatous damage will not occur. Therefore, the target pressure in an individual patient with glaucoma needs to be periodically reassessed and changed in the light of diurnal variations in IOP, visual field defects and changes in ONH.

The reduction of intraocular pressure can be obtained medically, surgically or by a combination of the two. Medical therapy is generally preferred for open-angle glaucoma and it should be instituted as soon as the disease is diagnosed.

The drugs used act either by decreasing the rate of aqueous formation or by increasing the rate of aqueous outflow, or both.

Recently, two new concepts in glaucoma therapy are surfacing: (i) *to enhance the blood flow of ONH*, and (ii) *to protect the ganglion cells from early death (neuroprotection).* Presently, none of the available drugs has these beneficial effects. Therefore, the goal of current therapy is to lower the IOP. The medical treatment that achieves this goal with lowest risk and fewer side effects should be employed. The commonly used anti-glaucoma drugs are classified as follows.

OCULAR HYPOTENSIVE AGENTS

Cholinergic Drugs

Agonist
- Pilocarpine
- Carbachol

Adrenergic Drugs

Nonselective
- Epinephrine (adrenaline)
- Dipivalyl epinephrine

Selective
- Clonidine
- Apraclonidine hydrochloride
- Brimonidine

Beta-blockers

Nonselective
- Timolol maleate
- Levobunolol
- Carteolol
- Metipranolol

Selective
- Betaxolol

Prostaglandins

Latanoprost
Travoprost
Bimatoprost
Unoprostone

Carbonic Anhydrase Inhibitors

Systemic
- Acetazolamide
- Dichlorphenamide
- Methazolamide

Topical
- Dorzolamide
- Brinzolamide

Cholinergic Drugs

The cholinergic agents used are those having a direct parasympathomimetic effect resembling the action of acetylcholine at the receptor sites.

Pilocarpine is a parasympathomimetic drug which is currently less frequently used in open-angle glaucoma. It is used as drops in 0.50 to 6% solution. Pilocarpine can reduce the IOP by 15-25%. Pilocarpine pulls the scleral spur to tighten the trabecular meshwork and thus increases the outflow of aqueous thereby reducing the IOP.

The pressure lowering effect of pilocarpine begins within 20 minutes and reaches its peak in about 90 minutes and lasts for 4 hours. For a slow and sustained release of the drug, pilocarpine may be administered by ocuserts or in soaked hydrophilic contact lenses. Ocuserts are available as Pilo-20 system (1% solution) and Pilo-40 system (2 to 4% solution). They can be inserted either in the lower or upper fornix for a constant release of a steady concentration of the drug for 7 days. Similarly, pilocarpine gel (0.5" strip) can be applied in the lower fornix. The concentration and frequency of instillation of drug should be increased if the IOP does not normalize.

Carbachol has both direct and indirect actions. It is usually used in 1.5 to 3 percent concentration three times a day.

Practically all miotics produce side effects due to miosis which include diminished night vision, reduced visual acuity, particularly in the presence of axial lens opacities, myopia due to spasm of accommodation and generalized constriction of visual field. Occasionally, retinal detachment and rise of IOP (due to pupillary block) may occur.

Adrenergic Agonist

Adrenergic agonists are divided into selective and nonselective agents.

Nonselective Adrenergic Agonist

Epinephrine and dipivefrin increase the trabecular and the uveoscleral outflow. They also decrease the aqueous production. Nonselective adrenergics are replaced with a more effective selective alpha 2- adrenergic agonists.

Epinephrine (adrenaline) is a mixed alpha and beta agonist. The drug is used in 0.5%, 1% and 2% concentrations and administered twice daily. Side effects like ocular irritation, blepharoconjunctivitis, conjunctival pigmentation, precipitation of angle-closure glaucoma (due to mydriatic effect), cystoid macular edema, elevated blood pressure and cardiac arrhythmias may occur.

Dipivalyl epinephrine (dipivefrin) is a prodrug which is converted into epinephrine after absorption into the eye. It is used in 0.1% concentration twice daily. It is superior to epinephrine because of better corneal penetration, greater hypotensive effect (10 times greater than epinephrine) and fewer side effects.

Selective Alpha-2 Adrenergic Agonist

The mode of action of alpha-2 adrenergic agonist is not fully understood. It decreases the aqueous production and episcleral venous pressure and improves the aqueous humor outflow.

Clonidine hydrochloride is a selective alpha-2 adrenergic agonist. Topical clonidine 0.125-0.5% used thrice daily lowers the IOP by decreasing the aqueous humor production. It causes fall in blood pressure due to its central action.

Apraclonidine hydrochloride, a selective alpha-2 adrenergic agonist, is available as 0.5% ophthalmic solution. It is indicated for short-term adjunctive therapy especially in patients with maximally tolerated medical therapy and in diminishing the acute IOP rise following laser iridotomy, laser trabeculoplasty and laser capsulotomy.

Brimonidine is much more highly selective alpha-2 adrenergic agonist than apraclonidine. It lowers the IOP by decreasing the aqueous formation and increasing the uveoscleral outflow. The drug is used in two concentrations, 0.2 and 0.15%. The latter has been shown to be as effective as 0.2% but with fewer side effects. Systemic side effects of brimonidine include dry mouth, drowsiness and lethargy. It should be avoided in infants due to an increased risk of hypotension, seizures and apnea.

Beta-Adrenergic Antagonists or Beta-Blockers

Timolol maleate is a nonselective beta-blocker. Timolol maleate is used in 0.25 or 0.5% concentration and administered twice daily. As the drug does not affect the size of the pupil and accommodation, the patients of glaucoma with central nuclear sclerosis can use it. However, it should not be used in patients with bronchial asthma and cardiovascular problems because of possibility of inducing bronchial spasm and vascular hypotension. The local side effects include burning, corneal anesthesia and punctate keratitis. Timolol hemihydrate (0.5%) has similar actions as that of timolol maleate but is less expensive.

Levobunolol is also a nonselective β-blocker available in 0.25-0.5% concentrations. It reduces the intraocular pressure maximally between 2 and 6 hours after instillation. The hypotensive effect of levobunolol is comparable to that of timolol maleate. The drug should be used with caution in patients with cardiovascular and obstructive pulmonary disorders.

Carteolol hydrochloride (1%), a nonselective β-blocking agent with associated sympathomimetic activity, is effective in lowering the intraocular pressure maximally 4 hours after the instillation.

Metipranolol (0.3%) is also a nonselective β-blocker. The peak reduction in IOP occurs 2 hours after the instillation. The drug has adverse pulmonary and cardiac effects.

Betaxolol is a selective beta-blocker which blocks mainly the β_1-receptors. Topical betaxolol lowers the intraocular pressure by 15-20% and the peak reduction is noted within 2-3 hours after the instillation in normal and glaucomatous eyes. It is used in 0.25 or 0.5% concentration twice daily. Betaxolol is the topical beta-blocker of choice in patients with open-angle glaucoma associated with pulmonary problems. However, respiratory difficulties are noticed after the use of betaxolol in certain susceptible and high-risk patients.

Prostaglandins (Hypotensive Lipids)

Hypotensive lipids currently used in glaucoma include prostaglandin analogs (latanoprost and travoprost), bimatoprost and unoprostone isopropyl. All these agents act by increasing the aqueous outflow. Long-term use of these drugs causes iris pigmentation . Other side effects include conjunctival hyperemia, trichiasis, pigmentation of the eyelid skin, hair growth around the eye and exacerbation of cytoid macular edema and uveitis.

Latanoprost is a prodrug activated by esterase during its passage through the cornea. It lowers the IOP by 25-32% by increasing the aqueous outflow through the uveoscleral pathway. Latanoprost (0.005%) is used topically once a day preferably in night. It is as effective or even better

than timolol 0.5% in lowering the IOP. It has additive effect when combined with timolol.

Travoprost is hydrolyzed by corneal esterase . It works by increasing the uveoscleral outflow and thus reduces the IOP approximately by 25%. Travoprost is used in 0.004% strength once a day at night time.

Bimatoprost is a prostamide. It lowers the IOP by 27-33% by increasing the uveoscleral as well as the trabecular outflow. Bimatoprost is available in 0.03% concentration and used once a day at night time .

Unoprostone is a docosanoid derivative developed in Japan. It is less effective in lowering the IOP (13-18%) than latanoprost. However, it has almost no side effect except corneal toxicity. Unoprostone (0.15%) is used topically twice a day. It acts by increasing the uveoscleral outflow.

Carbonic Anhydrase Inhibitors

Carbonic anhydrase inhibitors (CAIs) decrease the aqueous humor formation by direct antagonist activity on carbonic anhydrase of the ciliary epithelium. Over 90% of the ciliary epithelial enzyme must be abolished to decrease the aqueous production for lowering IOP. CAIs may be administered systemically or topically.

Systemic Carbonic Anhydrase Inhibitors

Acetazolamide is a potent anti-glaucoma drug which reduces the intraocular pressure by decreasing the carbonic anhydrase dependent aqueous production. The drug reduces the aqueous formation up to 15-20% by decreasing the availability of bicarbonate. Acetazolamide is generally administered orally in doses of 250 mg four times a day in the management of the acute congestive glaucoma. Sustained action capsules of 500 mg (diamox sequels) have a prolonged effect. Parenteral acetazolamide can be administered in the dose of 5-10 mg per kg body weight.

Dichlorphenamide (Daranide) is a carbonic anhydrase inhibitor having a longer duration of action. It is administered 50 mg twelve hourly.

Methazolamide (Naptazane) gives good hypotensive effect in lower doses (25 mg twice a day) and does not cause systemic acidosis.

Carbonic anhydrase inhibitors are derived from sulfa drugs and may cause similar allergic reactions. CAIs should not be administered for a long duration as they cause acidosis, paresthesia, anorexia, nausea and vomiting. Renal colic, blood dyscrasias, dermatitis, neuropathy, lenticular myopia and retinal edema are other side effects of the drug.

Topical Carbonic Anhydrase Inhibitors

Dorzolamide hydrochloride (2%) is a topical carbonic anhydrase inhibitor administered 3 times daily. It is useful as an adjunct to β-blockers or miotics in unresponsive patients.

Brinzolamide (1%) is also a topical carbonic anhydrase inhibitor used 2-3 times daily. It can be used as an adjunct with topical beta-blockers.

Topical CAI inhibitors are sulfonamides, therefore, may cause adverse reactions in sulfa-sensitive patients. The ocular side effects include burning, stinging, blurred vision, induced myopia and superficial punctate keratitis.

The medical therapy should be continued in open-angle glaucoma as long as the deterioration does not occur in the visual acuity and visual fields. The surgical intervention is indicated in patients in whom medication fails to lower the IOP to the target level (30% reduction from the baseline pressure), development of allergy or toxicity to the drug and noncompliance on the part of the patient.

Surgery for POAG

Several operations have been devised to control the IOP in open-angle glaucoma. The surgical procedure of choice for open-angle glaucoma is

an operation which establishes a communication between the anterior chamber and the subconjunctival space and thus bypasses the obstructed trabecular meshwork.

Trabeculectomy is the most commonly performed operation. The surgical procedure of trabeculectomy is described in the chapter on *Operations upon the Eyeball and its Adnexa.*

The outcome of surgery is good only if the operation is undertaken before the raised intraocular pressure has caused serious damage to the optic nerve fibers. The visual prognosis is poor if the surgery is performed in the late stages of the disease.

Laser Trabeculoplasty

Besides medical therapy and surgery, argon or diode laser trabeculoplasty (LT) can be performed to control the intraocular pressure in patients with POAG. This procedure is not a substitute for medical therapy but may be an alternative to the filtration surgery; in most cases it can delay the surgical intervention.

Primary Angle-Closure Glaucoma

Primary angle-closure glaucoma (PACG) is also known as *narrow-angle glaucoma*. It is the most common form of glaucoma in the East Asian countries but occurs less frequently in the West.

Risk Factors for Developing PACG

1. Race: The prevalence of PACG over the age of 40 years varies in different races: mixed ethnic group, 0.1-0.6%; white, 0.1-0.2%, and black, 0.4-0.14%.
2. Age: PACG is uncommon under the age of 40 years. Its prevalence increases with advancing age.
3. Gender: PACG occurs 2-4 times more frequently in females than in males.
4. Family history: PACG is more common in first degree glaucoma relatives.
5. Personality: PACG is more common in highly anxious and sympatheticotonic persons.

Ocular Biometrics

The type of the eye predisposed to PACG has following characteristics:

1. The eye is small with short axial length and is usually hypermetropic.
2. The cornea has small diameter and radius of curvature.
3. The anterior chamber is shallow; most patients with PACG have 2.1 mm depth.
4. The lens is thick with increased anterior curvature.
5. The root of iris is inserted comparatively more anteriorly on the anterior surface of ciliary body and the angle of the anterior chamber is always narrow (Fig. 15.25).

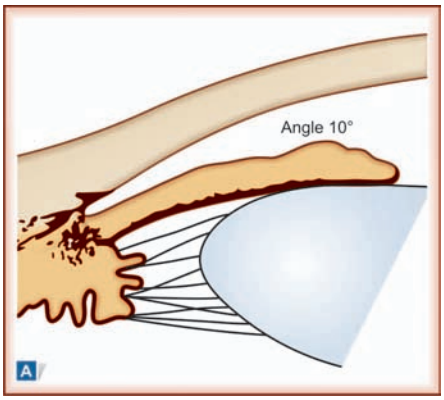


Fig. 15.25A: Diagrammatic representation of the angle of anterior chamber showing extremely narrow angle

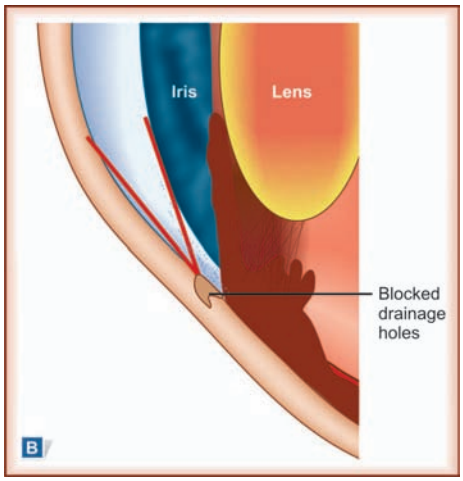


Fig. 15.25B: Narrow angle of the anterior chamber

Mechanism of Closure of the Angle of the Anterior Chamber

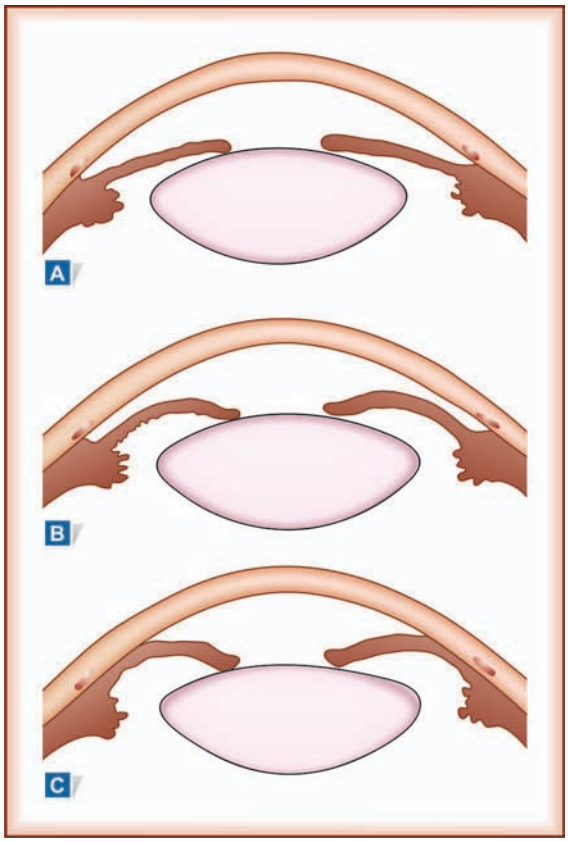
Every eye with narrow angle does not develop a glaucomatous attack as the intraocular pressure can be maintained within normal limits even if one-third circumference of the angle remains open. The mechanism of closure of angle of the anterior chamber varies considerably. In an eye with normal depth of the anterior chamber, the iris lies flatly in a transverse plane and its pupillary margin just touches the anterior surface of the lens. While in an eye predisposed to angle-closure glaucoma, the iris remains in close contact with the anterior surface of the lens with a considerable pressure from sphincter pupillae (Fig.15.26A). This contact embarrasses the circulation of aqueous from the posterior to the anterior chamber resulting in a relative pupillary block leading to a higher pressure in the posterior chamber. The peripheral iris becomes more flaccid and bows forwards, *iris bombé* (Fig. 15.26B). The iridotrabecular contact causes appositional angle-closure and obstruction to the aquous outflow (Fig. 15.26C). Long-standing iridotrabecular contact may form peripheral anterior synechiae (PAS).

The angle closure may also occur from crowding of the iris following dilatation of the pupil, or from the anteflexed ciliary body unassociated with pupillary block (as seen in the malignant glaucoma). Therefore, mydriatics must be used with caution in an eye with narrowangle and shallow anterior chamber.

Clinical Features

The clinical course of angle-closure glaucoma is divided into five stages. The disease may not progress from one stage to the other in an orderly manner.

1. Prodromal stage
2. Stage of constant instability
3. Acute congestive stage
4. Chronic angle closure, and
5. Absolute glaucoma.



Figs 15.26A to C: Mechanism of angle-closure glaucoma. A: Relative pupil block; B: Iris bombé; C: Iridotrabecular contact

Prodromal stage: This stage is marked by occasional transient attacks of raised intraocular pressure associated with colored halos due to corneal edema and headache. In spite of raised IOP (40-60 mm Hg), the eye with shallow anterior chamber remains white. Slit-lamp examination may reveal corneal edema and irregular anterior chamber depth owing to iris bombé. The attacks are usually precipitated by anxiety and overwork, and subside without any medication.

Stage of constant instability: The attacks of raised intraocular pressure are more frequent and occur with regularity. The diurnal variation of intra-ocular pressure occurs secondary to the vascular strangulation particularly in the late afternoon and evening. Subclinical episodes of raised IOP

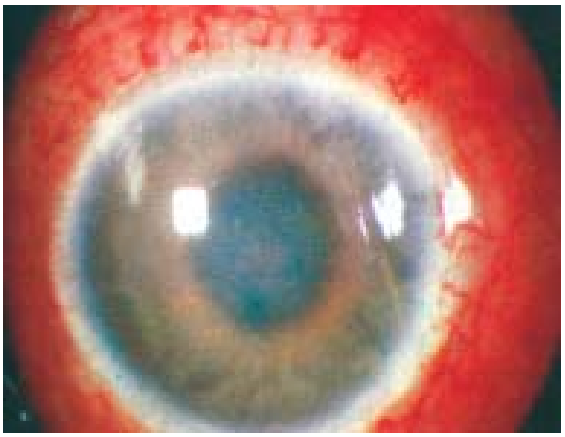


Fig. 15.27: Acute congestive glaucoma

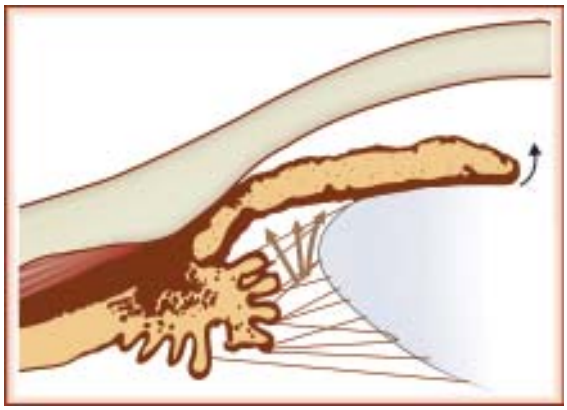


Fig. 15.28: Diagrammatic representation of the angle of anterior chamber showing angle closure

are often associated with slowly progressive closure of the angle of the anterior chamber. However, rest and comfortable sleep induce a quick fall in the ocular tension.

The dignosis of PACG during prodromal stage or stage of constant instability can be made with high index of suspicion and proper gonioscopy. The disease may pass on to the acute congestive or chronic stage if not treated. Laser iridectomy is the treatment of choice.

Acute congestive stage: An acute congestive attack is characterized by a sudden neuralgic pain, profound diminution of vision, intense ciliary congestion, corneal edema (Fig. 15.27), very shallow anterior chamber, complete closure of the angle of the anterior chamber, vertically dilated non-reacting pupil and markedly raised IOP.

An acute congestive attack occurs always with the closure of the angle by peripheral anterior synechiae and edematous and congested root of the iris and ciliary processes (Fig. 15.28). The changes in the iris are secondary to the vascular strangulation which results from the raised intraocular pressure.

A typical attack occurs unilaterally either in a darkened environment (causing dilatation of pupil) or following emotional crisis. Sometimes, both eyes are affected by an inadvertent use of a mydriatic drug.

Clinical Features

The classical symptoms include severe ocular pain, headache, blurred vision, rainbow-colored halos around the light, nausea and vomiting. The pain is due to stretching of the sensory nerves and radiates over the entire distribution of the fifth cranial nerve. The pain may induce nausea and vomiting and thus be mistaken for a bilious attack. There is a rapid and marked deterioration in the visual acuity of the affected eye and the vision may be reduced to hand movements. The halos (rainbow vision) are frequent due to corneal edema. Lacrimation and photophobia are common. The eye is congested and suffused due to congested episcleral and conjunctival vessels.

The lids and conjunctiva are edematous and the ciliary congestion is marked. The cornea is steamy and insensitive. The anterior chamber is extremely shallow. The aqueous shows the presence of flare due to ischemic damage to the uveal tissue resulting from the rise of IOP.

The iris appears discolored. Owing to pressure on the ciliary nerve and sphincter pupillae and

edema of the iris tissue, the pupil is dilated and vertically oval and may not react to light and accommodation. The iris ischemia may produce iris atrophy and cause permanently dilated and fixed pupil. It may lead to release of iris pigments and dusting of corneal endothelium. Opacities in the anterior lens cortex, glaukomflecken, may also develop as a result of ischemia.

The ocular tension is usually very high. The eyeball is tender. Gonioscopy often reveals a completely closed angle. However, compression gonioscopy can differentiate between a reversible and irreversible angle closure.

The fundus of the patient can be examined following instillation of glycerine drops which relieve corneal edema. The optic nerve head may be swollen, small hemorrhages on the disc, retinal vascular occlusion and spontaneous pulsations of the retinal artery may be seen. Glaucomatous cupping is a feature of long-standing untreated case.

The acute congestive attack may subside with prompt and aggressive treatment. However, the untreated case may pass into the chronic congestive stage. Rarely, an acute attack of glaucoma may terminate into absolute glaucoma wherein the eye is completely blind. Recurrences of acute attack are not uncommon. Each attack further closes the angle of the anterior chamber by forming peripheral anterior synechiae and leads to further deterioration of vision, constriction of the visual field and damage to the optic nerve fibers.

Chronic angle closure: This stage can develop either after acute angle closure or when the angle closes gradually and IOP rises slowly. A gradual asymptomatic angle closure is known as *creeping angle closure* in which a slow PAS formation develops circumferentially. Clinical course of chronic angle-closure glaucoma resembles that of POAG. It presents a few symptoms, moderate rise of IOP, glaucomatous optic neuropathy and characteristic visual field defects. The gonioscopic evaluation enables the ophthalmologist to differentiate between the two.

Stage of absolute glaucoma: In this stage, the eye becomes painful and blind. A chronic congestion is seen in the circumcorneal region and often the anterior ciliary vessels are dilated. The cornea is edematous and may have bullous (vesicles) or filamentary keratopathy; it is hazy and insensitive. The anterior chamber is very shallow. The iris may show atrophic patches. The pupil is dilated and does not react to light and accommodation. Ectropion of uveal pigments is frequent at the pupillary border. The IOP is very high and the eyeball is stony hard. The optic nerve head is deeply cupped. The sustained elevation of intraocular pressure causes weakening of the sclera and formation of ciliary staphyloma and equatorial staphyloma (Fig.15.29). Later, degenerative changes in the ciliary body result in decreased aqueous formation which may normalize or decrease the tension. Shrinkage of the eyeball may occur due to marked hypotonia.

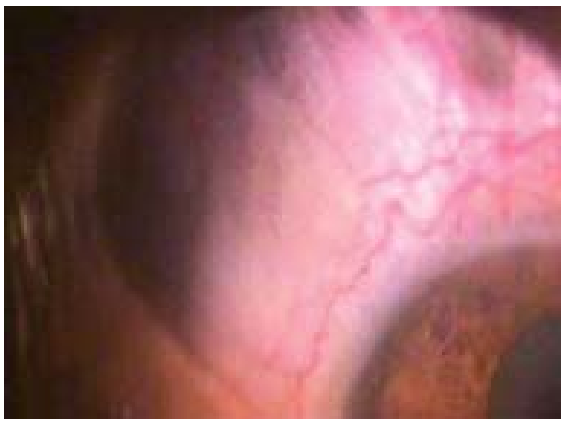


Fig. 15.29: Ciliary and equatorial staphylomas

Diagnosis

Angle of the anterior chamber: The diagnosis of angle-closure glaucoma in the prodromal stage is important. It may be emphasized that during the prodromal stage of angle-closure glaucoma

usually the intraocular pressure is not raised between the attacks, and visual acuity and visual field remain within normal limits. The eye appears clinically normal except for the narrowness of the angle. The dignosis requires a clinical judgement and an accurate assessment of the angle of the anterior chamber. On repeated gonioscopy it must be assessed whether the narrow angle has an appositional closure.

Colored halos: The history of seeing colored halos in a highly anxious woman patient in her fifties should always arouse the suspicion of the disease. The colored halos of glaucoma are due to corneal edema caused by raised intraocular pressure and must be differentiated from the halos found in acute purulent conjunctivitis and early immature cataract. The halos associated with conjunctivitis can be eliminated by the irrigation of discharge from the conjunctival sac. Fincham's test can differentiate between the halos of glaucoma and immature cataract. The test comprises a stenopeic slit which is passed before the eye across the line of vision. The glaucomatous halo does not alter, while the lenticular halo is broken up into segments with the passage of the slit (Fig. 15.30).

Anterior chamber: The eye of a patient with PACG almost always has a shallow anterior chamber which can be determined by slit-lamp. Shallowness of the anterior chamber is almost invariably accompanied by narrowness of the angle which can be confirmed on gonioscopy. During prodromal attack, the angle becomes narrow owing to accentuation of the physiological *iris bombé*. But permanent adhesions between the root of the iris and the posterior surface of the cornea, known as *peripheral anterior synechiae*, do not develop. However, since the upper angle is relatively narrow, peripheral anterior synechiae may be formed here in subsequent attacks. They gradually spread around the periphery and the eye is likely to develop an acute congestive attack when three-fourth of the circumference of the angle is occluded.

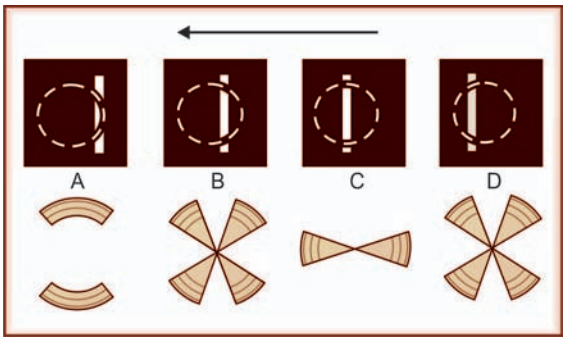


Fig. 15.30: Emslay-Fincham test: when a stenopeic slit is passed before the eye, the lenticular halos change their pattern as per the exposed lens fibers. A: Horizontal radial fibers, B and D: Oblique fibers, C: Vertical fibers

Provocative tests: As stated, the recording of IOP in prodromal stage is nonconclusive. Certain provocative tests are designed to study the trend of IOP. They are based on inducing transient pupillary dilatation which further narrows the angle of the anterior chamber in an eye predisposed to angle-closure glaucoma. The prone darkroom and mydriatic tests can be used in the diagnosis of angle-closure glaucoma.

Prone darkroom test: In the prone position an angle-closure may develop due to the pupillary block associated with a slight anterior shift of the lens. The baseline IOP is recorded and the suspect is placed in the prone position for 60 minutes. A rise of 8 mm Hg or more of intraocular pressure is considered to be a positive test.

Mydriatic test: The baseline IOP is recorded, then the pupil is dilated with a weak mydriatic such as phenylephrine (2%). Measurements of IOP is carried out at 30 minutes interval for two hours. A difference of 8 mm Hg between the initial reading and the peak rise following pupillary dilatation suggests the possibility of angle-closure glaucoma. The precipitation of acute congestive attack may follow a mydriatic test, hence, the patient must remain under observation until the pupil attains its normal size.

Predictive values of the provocative tests have not been demonstrated in the recent studies. When the provocative tests are negative the presence of the PACG can not be excluded nor can any assurance be given that an acute congestive attack will not ensue in the future. Suspected cases of narrow angle glaucoma should be advised to report for regular follow-up examination. The examination of the other eye may provide important clues to the diagnosis as the disease is often bilateral.

Treatment

Primary angle-closure glaucoma carries an excellent prognosis if treated in the prodromal stage. It is often managed surgically and the medical treatment is usually limited to the preoperative reduction of intraocular pressure.

Pilocarpine: During the prodromal stage and stage of constant instability halos appear at a particular hour of the day (mostly late afternoon or evening), coinciding with the peak rise of IOP. It is advisable, therefore, to instill pilocarpine 0.5% to 1% half an hour before the appearance of halos. Pilocarpine causes miosis and relieves crowding of the iris at the angle of the anterior chamber and prevents the formation of peripheral anterior synechiae.

Laser iridectomy: The laser iridectomy is the treatment of choice for the management of early stages of PACG. A peripheral iridectomy performed before the development of PAS virtually cures the condition as it establishes a free communication between anterior and posterior chambers and abolishes iris bombé. The operation is simple and is practically without any risk or complication.

Treatment of Acute Congestive Stage

The medical treatment of acute congestive angle-closure glaucoma is aimed at preparing the patient for laser iridectomy. The treatment must reduce the IOP rapidly to prevent damage to vital eye structures.

Incisional surgery is generally avoided during an acute congestive attack of glaucoma because of difficulties of operation on a suffused eye and the dangers of opening the globe having a very high pressure.

To quell an acute attack both topical and systemic hypotensive agents should be used. The administration of topical corticosteroids 3-4 times a day reduces the accompanying inflammation in the eye. The mild attacks of acute angle-closure glaucoma may be broken by 1-2 % pilocarpine eye drop 4-6 times a day, which induces miosis and pulls the iris away from the trabeculum. When the IOP is more than 50 mm Hg, the sphincter pupillae is ischemic and may not respond to pilocarpine therapy. A combination of topical timolol maleate and brimonidine or topical and oral CAIs should be used. When necessary a hyperosmotic agent like oral glycerol (1.5 g/kg body weight) or IV mannitol (1 g/kg body weight) should be administered. Besides medication, globe compression and compression gonioscopy have been recommended to reduce the IOP.

Hyperosmotic Agents

Hyperosmotic agents are of great value in controlling the acute phase of primary angle-closure glaucoma. They act by drawing the water out of the eye and reduce the IOP. The commonly used hyperosmotic agents are glycerol, mannitol, urea and isosorbide.

Glycerol

Glycerol is a syrupy liquid with a sweet taste. It is given orally in a dose of 1.5 gm/kg body weight as a 50% solution with lemon to improve the flavor. The maximal hypotensive effect of glycerol starts within one hour and lasts for nearly 3 hours. Nausea, vomiting and headache are common side effects of glycerol therapy. The drug should be administered in diabetics with caution.

Mannitol

Mannitol is administered intravenously as 20% solution in water in the dose of 1 to 2 gm/kg body weight over a period of 30 to 40 minutes. It penetrates the eye poorly, therefore, reduces the intraocular pressure effectively within 30 minutes and its effect lasts for about 4 hours. The drug is contraindicated in patients with renal disease and should be used with caution in congestive heart failure.

Urea

Urea is used intravenously as a 30% solution in 10% inert sugar in the dose of 1 gm/kg body weight. Its use is contraindicated in patients with impaired renal function.

Isosorbide

Isosorbide is administered orally in the dose of 1 to 2 gm/kg body weight. It has a minty flavor and is free of nausea. The drug can be even given to diabetic patients.

To allay pain in the acute congestive stage of angle-closure glaucoma, it is necessary to administer analgesics. A peribulbar injection of 1 ml of 2% xylocaine with adrenaline 1 in 10000 gives great relief owing to its hypotensive and anesthetic effects.

Surgery

Once the acute attack is broken and cornea regains clarity, surgery must be performed. The nature of operation depends on the gonioscopic appearance of the angle of the anterior chamber. In the absence of peripheral anterior synechia, a laser iridotomy is the most preferred surgery. But if goniosynechiae are extensive, a filtration operation is indicated.

Fellow Eye

The untreated fellow eye has a 40-80% chance of developing acute attack of PACG in 5-10 years period. Therefore, prophylactic iridectomy should be performed in the eye unless the angle is clearly nonoccludable.

Treatment of Chronic Congestive Glaucoma

As the diagnosis of chronic congestive glaucoma is established, an operation is warranted. The development of peripheral anterior synechiae in the filtration angle does not allow the iris to fall away from the posterior surface of the cornea despite a peripheral iridectomy. In these cases a filtration operation should be performed.

Treatment of Absolute Glaucoma

In absolute glaucoma, the IOP can be lowered by cyclodestructive procedures. If the eye is painful, enucleation is indicated. In such eyes the pain may be relieved temporarily by retrobulbar injection of 1 ml of 2% xylocaine followed seven minutes later by 1 ml of 80% alcohol.

PACG with Plateau Iris

Primary angle-closure glaucoma, although uncommon, may be caused by anteriorly positioned ciliary processes which push the peripheral iris anteriorly and close the angle of the anterior chamber. Plateau iris syndrome can be diagnosed by ultrasound biomicroscopy. The glaucoma can be managed by miotics or laser iridoplasty (gonioplasty).

SECONDARY GLAUCOMAS

Classification

The secondary glaucoma can be divided into following categories:

1. Secondary glaucoma with open angle
2. Secondary glaucoma with angle closure:
   a. Angle-closure with pupillary block
   b. Angle-closure without pupillary block.

Some types of secondary glaucomas, such as uveitic, lens induced and traumatic, may present with open angle as well as angle closure. In contrast to the primary, the etiology of the secondary glaucoma is fairly known. Several factors such as inflammation, neoplasia, trauma, disorders of the lens and anomalies of the angle of anterior chamber may cause the secondary glaucoma. Generally, the disease process obstructs the trabecular meshwork or the pupil and lead to rise in the intraocular pressure. Common types of secondary glaucomas are described below.

Glaucoma due to Uveitis

The intraocular pressure may rise both in the acute and the chronic anterior uveitis. IOP in acute anterior uveitis may be raised due to (i) trabecular obstruction by inflammatory cells and debris, (ii) inflammation of the trabecular meshwork (trabeculitis) which results in reduction of intertrabecular spaces, (iii) trabecular meshwork endothelial cells dysfunction, and (iv) breakdown of the blood aqueous barrier resulting in plasmoid aqueous.

The presence of KPs, aqueous cells, miotic pupil, PAS, iris bombé and raised IOP is diagnostic of uveitic glaucoma. The secondary glaucoma in chronic anterior uveitis occurs either due to seclusio pupillae (total ring synechia), occlusio pupillae or extensive peripheral anterior synechiae.

Secondary angle-closure can also occur due to uveal effusion, exudative retinal detachment and choroidal effusion followed by forward displacement of the iris-lens diaphragm.

The condition is managed by oral and topical CAIs and corticosteroids. The use of miotics is contraindicated as they perpetuate the inflammatory phenomenon and facilitate synechia formation.

The medical treatment of the condition is not effective. The pupillary block should be relieved by a peripheral iridectomy and the presence of extensive PAS warrants filtration surgery, trabeculectomy.

Secondary glaucoma is also found in Fuchs heterochromic cyclitis and glaucomatocyclitic crisis. They are described in the chapter on *Diseases of the uveal tract*.

Phacogenic Glaucoma

The lens may cause secondary glaucoma in a number of ways.

Phacomorphic Glaucoma

The lens may cause both open-angle and angle-closure secondary glaucomas. The PACG occurs in an eye predisposed to angle closure but phacomorphic glaucoma can occur in eyes not susceptible to closure. The process of development of phacomorphic glaucoma is much more rapid and is precipitated by swelling of the lens during intumescent stage of cataract and development of pupillary block.

The management of phacomorphic glaucoma includes oral CAIs, laser iridectomy and extraction of lens when eye becomes quiet.

Phacolytic Glaucoma

Phacolytic glaucoma is usually associated with mature or hypermature cataract and it occurs due to the leakage of lens proteins through an opening in the capsule. The leaked denatured lens proteins are engulfed by macrophages which subsequently block the trabecular pores. An acute or subacute rise of intraocular pressure causes pain and ciliary injection. The characteristic signs are microcystic corneal edema, marked cells and flare in the anterior chamber without KPs, cellular debris or clumps of protein in the anterior chamber, hypermature cataract and elevated IOP.

The extraction of lens is the only possible treatment, but it should be done after reducing the intraocular pressure by hyperosmotic agents.

Phacoanaphylactic Uveitis and Glaucoma

The hypersensitivity to patient's own lens protein induces an inflammatory reaction in the eye. In the event of penetrating ocular trauma or following extracapsular cataract extraction, a severe phacoanaphylactic reaction to the lens matter develops.

It is characterized by a moderate granulomatous anterior uveitis (with KPs, that distinguishes it from phacolytic glaucoma), lens matter in the anterior chamber, synechiae formation, vitritis and raised IOP.

The rise in intraocular pressure is due to the obstruction of trabecular meshwork by particulate matter. The condition must be treated by mydriatics and local and systemic corticosteroids. The lens matter should be aspirated or removed.

Glaucoma Associated with Dislocated Lens

An anteriorly dislocated lens may block the outflow of aqueous humor at the pupil and cause acute congestive glaucoma.

A pupillary block glaucoma occurs when the lens is small and spherical (microspherophakia) as seen in Weill-Marchesani syndrome.

Dislocation of the lens into the anterior chamber is an emergency as it causes a sharp rise in intraocular pressure and may permanently damage the corneal endothelium. Early surgery is warranted.

In pupillary block glaucoma, a mydriatic or peripheral iridectomy/laser iridectomy relieves the IOP. Miotics are contraindicated as they make the condition worse. Lens extraction is usually indicated to restore the vision and prevent the pupillary block.

Secondary glaucoma often occurs following dislocation of the lens into the vitreous cavity. It usually induces cyclitis and the inflammatory cells or degenerated lens material may block the trabecular meshwork causing rise of intraocular pressure.

Pseudoexfoliation Glaucoma (Exfoliation Syndrome)

More than 50% cases of pseudoexfoliation syndrome may develop open-angle glaucoma. A combination of pseudoexfoliation and glaucoma is known as *glaucoma capsulare.* Pseudoexfoliation is a basement membrane disorder of unknown etiology. The development of glaucoma largely depends on the extent and rapidity by which the fibrillogranular material and pigments accumulate in the trabecular meshwork and obstruct the outflow channels.

The dandruff-like material is deposited on the pupillary border of the iris and on the anterior lens capsule (except the central zone) (Fig. 15.31). In most cases the angle of the anterior chamber becomes narrow due to anterior movement of iris-lens diaphragm. The pigments are seen arranged in a linear fashion anterior to Schwalbe's line *(Sampaolesi's line*). Phacodonesis or subluxation of the lens may occur due to looseness of the zonule. The presence of greater pigmentation of trabecular meshwork and monocular involvement distinguishes exfoliation glaucoma from POAG.

The management of glaucoma associated with exfoliation is more difficult than the open-angle glaucoma without exfoliation. Laser trabeculo-

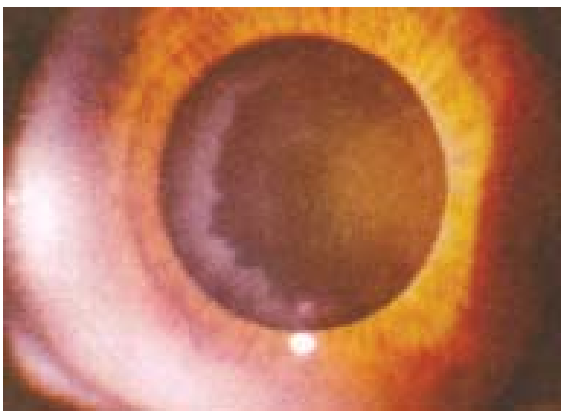


Fig. 15.31: Pseudoexfoliation of anterior capsule (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

plasty can be effective. The non-responsive cases can be dealt with trabeculectomy but post-operative intraocular reaction is quite common.

Traumatic Glaucoma

Traumatic or Angle-recession Glaucoma

A blunt injury to the eye can cause a tear in the anterior face of the ciliary body and recession of the angle of the anterior chamber. It is marked by retrodisplacement of the iris root. The glaucoma due to recession of the angle of the anterior chamber is usually chronic, unilateral and secondary open angle. The classical gonioscopy findings include broad angle recess, torn iris processes, white glistening scleral spur and localized depression of the trabecular meshwork.

An early onset glaucoma in the recession of the angle is due to hyphema, but late onset glaucoma is probably due to fibrosis of trabecular meshwork, extensive PAS and hyaline membrane formation.

A moderate rise of IOP following recession of the angle can be managed by miotic, epinephrine and/or carbonic anhydrase inhibitors. In severe cases, medical therapy is ineffective and a filtering operation becomes necessary.

Glaucoma due to Penetrating Injuries

Penetrating injury to the eye is a common cause of secondary glaucoma. Adherent leukoma (Fig. 15.32), pupillary block by dislocated swollen lens, traumatic uveitis and persistent hyphema are other modes of secondary glaucoma due to ocular injuries.

Proper repair of ocular wound should be performed. The pupillary block can be relieved by iridectomy or sphincterotomy. Mydriatic, antibiotic and corticosteroids should be used to prevent synechiae formation and infection.

Glaucoma Associated with Intraocular Hemorrhage

Hyphema

Traumatic hyphema (Fig. 15.33) is a common cause of secondary glaucoma. Secondary glaucoma is more frequent following recurrent bleeding. In general, larger the hyphema higher is the rise of IOP. The rise of IOP occurs as a result of obstruction of trabecular meshwork by hemorrhagic debris, fibrin, and lysed RBCs.

Hemolytic and Ghost Cell Glaucoma

Hemolytic and ghost cell glaucoma develop after vitreous hemorrhage. In hemolytic glaucoma red-tinged cells float in the anterior chamber and

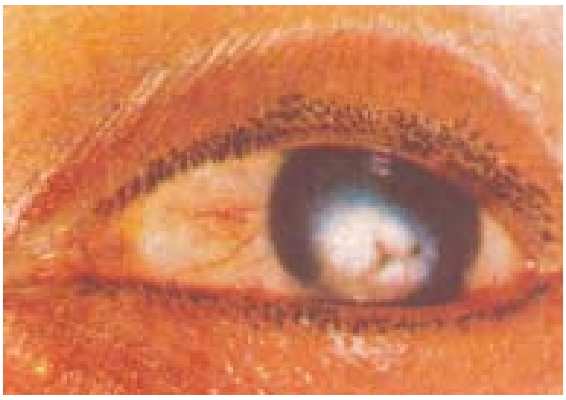


Fig. 15.32: Post-traumatic glaucoma

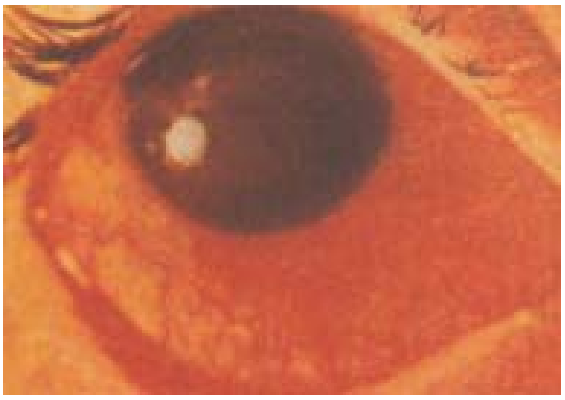


Fig. 15.33: Secondary glaucoma: subconjunctival hemorrhage and hyphema

macrophages filled with hemoglobin block the trabecular outflow channels. Within 1-3 months of vitreous hemorrhage, the red blood cells degenerate into ghost cells when hemoglobin leaks out. The ghost cells are spherical, 4 to 6 microns in diameter, hollow in appearance, khaki colored, and less pliable. Because they are rigid, they block the trabecular meshwork and produce *ghost cell glaucoma*. This type of glaucoma is common in aphakic eyes.

Aminocaproic acid, an antifibrinolytic agent, is given in a dose of 100 mg per kg body weight orally, 6 hourly for 5 days to prevent secondary hemorrhages. Medical therapy with aqueous suppressants can be effective. Some patients may need anterior chamber irrigation, pars plana vitrectomy and filtering surgery to control the elevated IOP.

Postoperative Glaucoma

Aphakic and Pseudophakic Glaucomas

Postoperative pupillary block may develop due to herniation of an intact face of vitreous in aphakia. Pupillary block can also occur following anterior chamber or posterior chamber intraocular lens (IOL) implantation. Capsular block, though uncommon, results from viscoelastic in the capsular bag, which pushes the posterior chamber IOL anteriorly causing closure of the angle of the anterior chamber. Pupillary block can be managed by multiple laser iridectomies.

Flat Anterior Chamber Glaucoma

A persistent postoperative flat anterior chamber often results in synechial closure of the angle of the anterior chamber and rise in IOP. If the anterior hyaloid or IOL is in the contact with the cornea, the anterior chamber should be reformed without any delay to prevent corneal endothelial damage.

Epithelial and Fibrous Downgrowth

Epithelial and fibrous downgrowth causes intractable secondary glaucoma. Postoperative wound dehiscence and delayed wound closure facilitate epithelial or fibrous downgrowth in the anterior chamber.The epithelial growth appears as gray vascular membrane which invades the posterior surface of the cornea, iris and trabecular meshwork.

Radical excision of the growth with repair of the wound is recommended but in most cases prognosis remains poor.

Glaucoma Associated with Retinal Surgery

Scleral buckling with encircling band may cause angle-closure glaucoma. The buckle can compress the vortex veins thereby increasing the episcleral pressure and IOP. The injection of air and expansile gases and silicone oil may result in angle-closure glaucoma. The glaucoma can be managed by release of band, removal of expansile gas or silicone oil. Non-responding cases need filtering surgery.

Malignant and Ciliary Block Glaucoma

Malignant glaucoma can occur in eyes with open angle following cataract surgery. It results from anterior rotation of the ciliary body causing posterior misdirection of aqueous humor in the vitreous cavity; hence it is also called *aqueous misdirection* or *posterior aqueous diversion* syndrome.

Clinically the anterior chamber is flat with forward bulge of the lens or vitreous face and marked rise of IOP. The ciliary processes are rotated anteriorly and may be visualized through an iridectomy opening. Optically clear aqueous zone can be seen in the vitreous.

The medical management of malignant glaucoma is difficult. It includes intensive therapy with beta blockers, CAIs and hyperosmotic agents.

Argon laser photocoagulation of the ciliary processes and anterior vitrectomy combined with anterior chamber reformation are more definitive treatment options.

Glaucoma Associated wih Nonrhegmatogenous Retinal Detachment

Nonrhegmatogenous retinal detachment is caused by retinoblastoma, malignant melanoma, subchoroidal hemorrhage, choroidal effusion, HIV infection and subretinal neovascularization. The accumulation of subretinal fluid pushes the retina forward against the lens and usually leads to raised IOP.

Glaucoma Associated with Elevated Episcleral Venous Pressure

Episcleral venous pressure is one of the important factors in the regulation of IOP. Normally the episcleral pressure ranges between 8 and 10 mm Hg. It may be raised in retrobulbar tumors, thyroid ophthalmopathy, superior vena cava syndrome, Sturge-Weber syndrome and arteriovenous fistula. Patients are symptom-free or present with chronic red eye. Tortuous and dilated episcleral vessels, raised IOP and blood in Schlemm's canal on gonioscopy are characteristic features. Aqueous suppressant therapy is effective, filtering surgery reduces IOP but may be complicated by suprachoroidal hemorrhage.

Neovascular Glaucoma

Neovascular glaucoma (NVG) is characterized by the formation of new vessels on the surface of the iris, *rubeosis iridis* (Fig. 15.34), and trabecular meshwork associated with raised intraocular pressure.

It is caused by central retinal vein occlusion, diabetic retinopathy and ocular ischemic syndrome. Sickle cell retinopathy, Eales' disease, longstanding retinal detachment, and intraocular tumors are the other causes. NVG is often induced by retinal ischemia or ocular inflammation. The ischemic retina releases a vasoformative substance which induces neovascularization of the anterior segment.

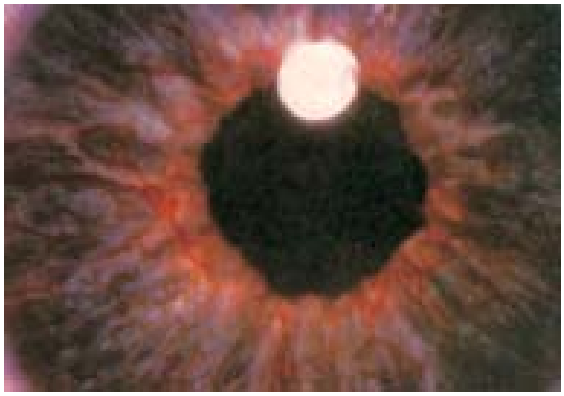


Fig. 15.34: Neovascular glaucoma associated with rubeosis iridis (*Courtesy:* Dr T Perkins, Madison)

The clinical features include an acute rise of IOP, ciliary injection, corneal edema, severe pain and markedly reduced vision. Gonioscopically, a fibrovascular membrane covering the trabecular meshwork is demonstrated. Recurrent hyphema often complicates the picture

The treatment of neovascular glaucoma is not effective. Topical beta blockers, alpha-2 adrenergic agonist, CAIs, corticosteroids and cycloplegic may reduce IOP as well as inflammation before surgery. Early neovascularization can be treated by panretinal photocoagulation and direct treatment of angle vessels with argon laser. Routine filtration surgery often fails to reduce the pressure, but Ahmed, Molteno or Krupin-Denver valve implantation, that drains aqueous into the subconjunctival space, is effective. If this fails, a cyclodestructive procedure may help reduce IOP and relieve pain.

Glaucoma Associated with Intraocular Tumors

Intraocular tumors like uveal melanomas and retinoblastoma may cause secondary glaucoma.

The tumor may induce rise in intraocular pressure in the following ways.

1. The rapidly growing tumor may push the lens-iris diaphragm forward thus causing angle-closure glaucoma.
2. An obstruction of trabecular meshwork may occur by tumor cells, macrophages containing tumor cells or inflammatory cells and debris.
3. Intraocular tumors may induce neovascularization of the iris and the angle of the anterior chamber, thus cause neovascular glaucoma.
4. Sometimes, the tumor is so situated as to press upon the vortex veins and impede the venous outflow from the eye resulting in secondary glaucoma.
5. Primary or metastatic tumors of the ciliary body may directly invade the angle of the anterior chamber.

Pigmentary Glaucoma

Raised IOP, mid-peripheral iris pigment atrophy and dispersion of pigments on the corneal endothelium in a vertical spindle pattern (Krukenberg spindle), trabecular meshwork, Schwalbe's line, iris surface, and lens equator characterize the pigmentary glaucoma.

Pigmentary glaucoma occurs in third to fifth decades. It affects mostly myopic males.The mechanism of pigment dispersion is not known. The *theory of reverse pupillary block* suggests that iris acts as a valve resulting in higher pressure in the anterior chamber than the posterior chamber causing posterior bowing of the iris. Pigment granules shedding from the iris occurs due to rubbing of the posterior surface of iris with the zonule. The released melanin granules block the trabecular meshwork leading to the rise of IOP. Gonioscopy may show a pigmentary line along the Schwalbe's line (Sampaolesi's line).

Miotics are effective in reducing the IOP. Argon laser trabeculoplasty and laser iridotomy may minimize the reverse pupillary block, if present. Filtration surgery is often successful in reducing the intraocular pressure.

Steroid Induced Glaucoma

The topically administered steroid drops and periocular or systemic corticosteroids cause a marked elevation of IOP in nearly 5% of the general population and moderate rise in 35%. The response of IOP to topical corticosteroids instillation is genetically determined. The Myocilin gene of primary open-angle glaucoma and that controlling corticosteroid responsiveness are closely related. The rise of IOP may occur due to an increased resistance to aqueous outflow in the trabecular meshwork. Deposition of mucopolysaccharides in the trabeculum, tightening of the lysosomal membrane and an increased aqueous humor formation are the probable mechanisms of steroid-induced glaucoma. Medrysone, loteprednol and flurometholone have a low potency for inducing ocular hypertension. It is, therefore, nondesirable to use potent water soluble corticosteroids (dexamethasone or betamethasone) for minor eye ailments for prolonged periods.

The withdrawal of topical corticosteroids lowers the pressure, however, some patients need treatment with topical β-blockers and systemic acetazolamide.

Epidemic Dropsy Glaucoma or Toxic Glaucoma

Toxic glaucoma may be found in patients of epidemic dropsy and is characterized by headache, colored halos, normal or deep anterior chamber, an open angle of the anterior chamber and marked elevation of intraocular pressure associated with generalized edema of the body.

The epidemic dropsy glaucoma is non-congestive in nature and is caused by the toxic action of *sanguinarine*, an active alkaloid in the seeds of *Argemone mexicana*. The glaucoma develops following consumption of mustard oil adulterated

with the oil of *Argemone mexicana*. Sanguinarine causes generalized capillary dilatation and increased formation of aqueous humor resulting in marked rise in IOP.

The signs and symptoms of epidemic dropsy glaucoma simulate open-angle glaucoma. Stoppage of consumption of adulterated mustard oil and administration of carbonic anhydrase inhibitor normalize the IOP. Refractory cases may need filtration operation.

Secondary glaucomas may also develop in a number of other clinical conditions such as iridocorneal endothelial syndrome (Chandler syndrome, essential iris atrophy, Cogan-Reese syndrome), nanophthalmos, retinopathy of prematurity, Fuchs hetrochromic iridocyclitis and glaucomatocyclitic crisis.

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CHAPTER

16 Diseases of the Lens

ANATOMY

The lens of the eye is a transparent biconvex avascular structure. It is suspended between the iris and the vitreous by the zonule which connects it with the ciliary body. It is surrounded by an elastic capsule which is a semipermeable membrane. The posterior surface of the lens is more curved than the anterior. The radii of curvature of anterior and posterior surfaces of the lens are 10 mm and 6 mm respectively. The centers of anterior and posterior surfaces are known as *anterior pole* and *posterior pole*, respectively. The consistency of the superficial part (*cortex*) of the lens is softer than the central (*nucleus*). The refractive index of lens is 1.39 and its diopteric power is approximately 17.75. The lens continues to grow throughout life and relative thickness of the cortex increases with age.

The lens consists of 3 parts (Fig. 16.1):

1. The lens capsule
2. The lens epithelium, and
3. The lens fibers.

The *lens capsule* is a transparent homogeneous and highly elastic envelope. The capsule is thicker in front than behind, the thickness being greater towards the equator, just anterior to the attachment of suspensory ligament, than at the pole. It is secreted by the lens epithelium.

The *lens epithelium* is a single layer of cubical cells that forms the anterior subcapsular epithelium. The posterior epithelial cells elongate to form the lens fibers in early embryonic life, hence the epithelium is not present posteriorly. The cells of epithelium are metabolically active and generate adenosine triphosphate (ATP) to meet the energy demand of the lens. The cells show high mitotic activity and form new cells which migrate towards the equator. The lens epithelial cells continue to divide and develop into the lens fibers.

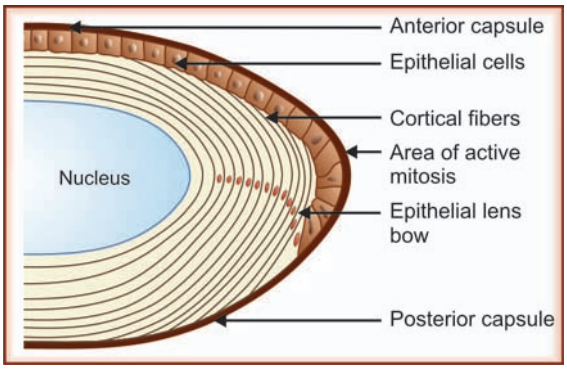


Fig. 16.1: Parts of lens

The *lens fibers* develop from the lens epithelial cells that continue to divide and get elongated and transformed into lens fibers. They are mainly composed of proteins called crystallins.

The fibers formed earlier lie in the deeper plane (nucleus of the lens), the newer ones occupy a more superficial plane. The fibers of embryonic nucleus meet around Y-shaped sutures. Surrounding the embryonic nucleus lies the fetal nucleus

corresponding to the lens at birth. The successive nuclear zones are infantile and adult nuclei (Fig. 16.2). The most peripheral part of the lens consists of cortex (young lens fibers) and the lens capsule (See Fig. 9.26). Each fiber starts anteriorly and ends posteriorly. Suture lines, formed by the end-to-end joining of these fibers, appear Y-shaped when viewed on a slit-lamp. The 'Y' is erect anteriorly and inverted posteriorly. There are more than 2000 fibers in an adult lens.

The lens shows changes with age. It is spherical, transparent and soft in infants. In adults, lens is firm, transparent and avascular. The adult lens measures 5 mm anteroposteriorly and 9 mm equatorially and weighs about 255 mg. In old age, it is of amber color, firmer in consistency and more flat on both the surfaces.

Zonular fibers or *suspensory ligament of lens* originate from basal laminae of the nonpigmented epithelium of ciliary body. The zonular fibers get inserted onto the anterior and the posterior lens capsule in a continuous fashion. The fibers hold the lens in position and enable the ciliary muscle to act during accommodation.

There are no blood vessels and nerves in the lens.

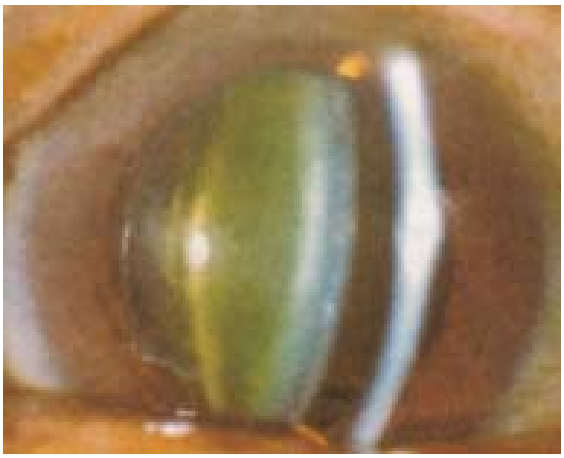


Fig. 16.2: Slit-lamp photograph of lens with various nuclei (*Courtesy*: Mr S Kanagami, Tokyo)

PHYSIOLOGY

The lens epithelium has the highest metabolic rate within the lens, it utilizes oxygen and glucose for protein synthesis and transport of electrolytes, carbohydrates and amino acids to the lens fibers. The lens maintains its transparency by maintenance of water and electrolyte balance. Normal lens contains approximately 66% water and 33% proteins. The lens cortex contains more water than the nucleus. The sodium concentration in lens is about 20 mM and potassium about 100 mM. The lens has higher levels of potassium ions and amino acids and lower levels of sodium and chlorine ions and water than the surrounding aqueous and vitreous humors. The electrolyte balance between inside and outside of the lens is maintained by selective permeability of the lens cell membrane and the activity of the sodium pump situated in the cell membrane of the lens epithelium and the lens fiber. The sodium pump acts by pumping sodium ions out and taking potassium ions in. This function depends on breakdown of ATP and is regulated by enzymes Na^+- K^+-ATPase. The combination of active transport and membrane permeability is known as *pump-leak system* of the lens. The lens epithelium is the primary site for active transport in the lens. The sodium is pumped across the anterior surface of the lens into the aqueous humor and potassium moves from the aqueous into the lens. At the posterior surface of the lens a passive diffusion of the solutes occurs. This arrangement results in a sodium-potassium gradient across the lens; potassium being higher at the front and sodium being higher at the back of the lens.

Normally the intracellular level of calcium in the lens is about 30 mM while the extracellular calcium level is close to 2 mM. The calcium pump maintains this gradient and is regulated by the enzyme Ca^{++}-activated ATPase. Loss of calcium homeostasis can derange the lens metabolism.

The sodium pump also helps in the active amino acids transport. However, glucose enters the lens by process of diffusion. The simple diffusion allows the waste products of the lens metabolism to leave the lens.

Lens derives its energy from carbohydrates and structural material from amino acids. As lens is an avascular structure, it has an overall low metabolic rate which is evident by the low rates of consumption of oxygen and utilization of glucose. The carbohydrate metabolism in the lens occurs by glycolysis, citric acid cycle, hexose-monophosphate shunt and sorbitol pathway. Amino acids and fatty acids are oxidized in the mitochondria of the lens epithelium via citric acid cycle.

DISEASES OF THE LENS

As the lens is an avascular structure it is incapable of becoming inflamed. However, degenerative changes in the lens are common and they often result in the partial or complete loss of transparency.

CATARACT

Any opacity in the lens or its capsule is known as *cataract*. Cataracts vary in degree of density and site and assume various forms. Clinically, cataract may be classified on the basis of morphology or underlying etiology.

Morphological Classification

Depending on the location and configuration of opacities, cataract can be classified as:

1. Capsular (anterior, posterior, bipolar)
2. Subcapsular (anterior, posterior)
3. Cortical
4. Supranuclear
5. Nuclear
6. Lamellar (zonular)
7. Sutural
8. Coralliform.

Etiological Classification

1. Congenital or developmental	Punctate, anterior polar, posterior polar, central nuclear, sutural, coralliform, zonular, coronary, membranous
2. Senile	Cortical, posterior subcapsular, and nuclear
3. Complicated	Uveitis, high myopia, retinitis pigmentosa, retinal detachment, glaucoma, ocular ischemia
4. Metabolic	Diabetes mellitus, tetany, galactosemia, Lowe's syndrome, Wilson's disease
5. Traumatic	Concussion injuries, penetrating injuries
6. Radiational	X-rays, gamma rays, neutrons, infrared, ultraviolet rays, microwave, laser radiations
7. Dermatogenic	Atopic dermatitis, Rothmund's syndrome, Werner's syndrome
8. Maternal infections	Congenital rubella, congenital toxoplasmosis, congenital cytomegalovirus disease, syphilis
9. Toxic	Corticosteroids, miotics, chlorpromazine
10. Cataract associated with systemic diseases	Dystrophia myotonica, Down's syndrome

Developmental Cataract

Developmental cataracts are usually present at birth *(congenital)* or may manifest after birth. The

etiology of many developmental cataracts is obscure. However, following factors play important role in the formation of developmental cataract.

1. *Heredity:* A strong hereditary predisposition is found in about 25% of all developmental cataracts.
2. *Intrauterine infections*: Rubella can cause cataract. Other infections such as cytomegalic inclusion disease, toxoplasmosis and syphilis can also lead to cataract formation.
3. *Radiation:* Exposure to radiation during pregnancy may cause cataract.
4. *Toxic agents:* Administration of corticosteroids or thalidomide during pregnancy has cataractogenic effect.
5. *Nutritional deficiency:* Deficiency states are often incriminated in the causation of zonular cataract.
6. *Miscellaneous causes:* Birth trauma, placental hemorrhage, endocrine dysfunction, and inborn errors of metabolism have been associated with developmental cataract.

The effects of infection, noxious agents, deficiency states or gene upon the developing lens are indistinguishable and mainly depend on the time of the insult. The most critical period in the development of the lens lies between 5th and 8th weeks of intrauterine life, when the cellular activity is maximal. Interference in the normal development during this period results in the formation of abnormal primary lens fibers leading to the development of central cataract. The involvement of secondary lens fibers during 8th to 16th weeks produces developmental cataract. Most developmental opacities are partial and stationary. The subsequent fibers are often normally formed and remain clear. Slight aberration in the development of lens fibers is common and, therefore, the lenses of most people show minute opacities especially when examined on slit-lamp under full mydriasis. It is advisable not to alarm such patients about their lens opacities as they rarely interfere with vision.

Developmental or congenital cataracts are broadly classified into two groups: *capsulolenticular cataract* wherein the capsule or the subcapsular region of the lens is involved, and *lenticular cataract* implicating the substance of the lens itself.

Several forms of developmental cataracts are found, the relatively common ones are described below.

Punctate Cataract

Punctate cataract is very frequent in occurrence and manifests as multiple, small opaque dots, scattered throughout the substance of the lens. On slit-lamp examination they appear as blue dots hence known as *blue-dot cataract* or *cataracta cerulea* (Fig. 16.3). A variant of punctate cataract is *cataracta centralis pulverulente*. It consists of fine white powdery dots within the embryonic and infantile nuclei. The punctate cataract is of little clinical significance.

Anterior Polar Cataract

The anterior polar cataract commonly occurs as a single or multiple opacities in the anterior part of the lens (Fig. 16.4). The remnants of the anterior

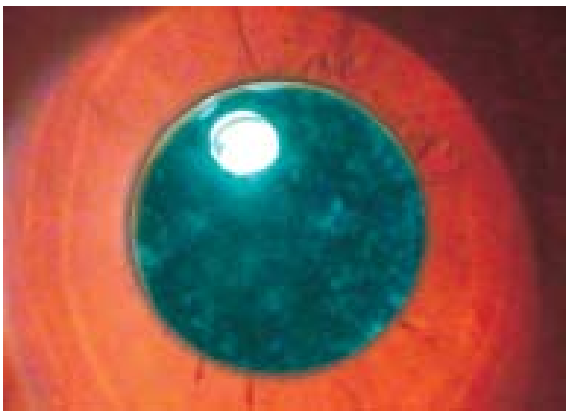


Fig. 16.3: Blue-dot punctate cataract

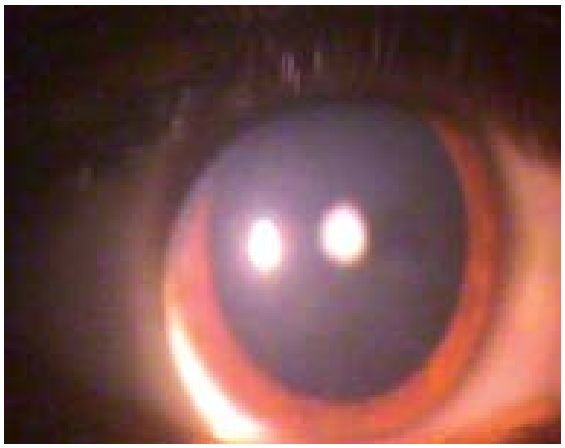


Fig. 16.4: Anterior polar cataract

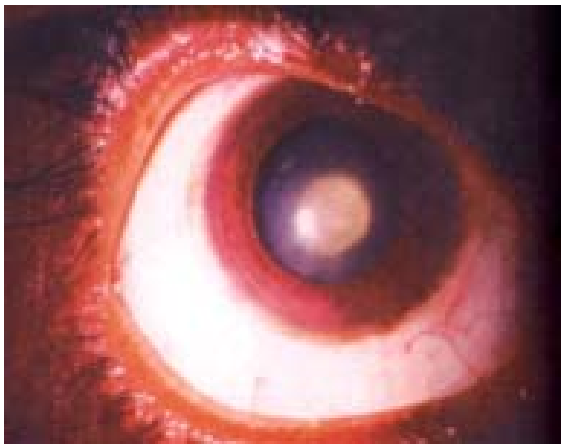


Fig. 16.5: Posterior polar cataract

(*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

vascular sheath are often found to be adherent to the opacities. The acquired form of anterior polar cataract occurs after perforation of a central corneal ulcer wherein part of the lens capsule comes in contact with perforated edges of the cornea. Occasionally the opaque lens capsule projects forwards into the anterior chamber like a pyramid, *anterior pyramidal cataract*. Later, normal transparent lens fibers grow between the capsular and cortical opacities, thus giving rise to a *reduplicated cataract*.

Posterior Polar Cataract

The posterior polar cataract is characterized by the presence of an opacity or opacities at the central posterior part of the lens and its capsule (Fig. 16.5). Slit-lamp examination reveals concentric rings around the central opacity (*onion-peel appearance*). The posterior polar cataract is usually associated with a thin posterior capsule or an occult posterior capsular defect.

The posterior polar cataract may be associated with persistence of the anterior tunica vasculosa lentis. *Mittendorf dot* (also known as *hyaloid corpuscle* as it represents the attachment of hyaloid vessel to the posterior capsule) is a small white dot attached to the lens capsule inferonasally. The dot may appear circular or rosette-shaped. It is usually stationary but the progressive form may appear which shows diffuse cortical opacities.

Central Nuclear Cataract

In the central nuclear cataract, the opacities are mostly confined to the embryonic nucleus (Fig. 16.6). It is due to the inhibition of development of the lens during the first three months of gestation. It is almost always bilateral. The opacity is heterogenous, either dense in the center and rare in the periphery or vice versa.

A progressive form of nuclear cataract is associated with the rubella (German measles) infection in the mother in which the entire lens may be opacified as the virus causes necrosis of the embryonic nucleus. Besides cataractous lens, other ocular changes

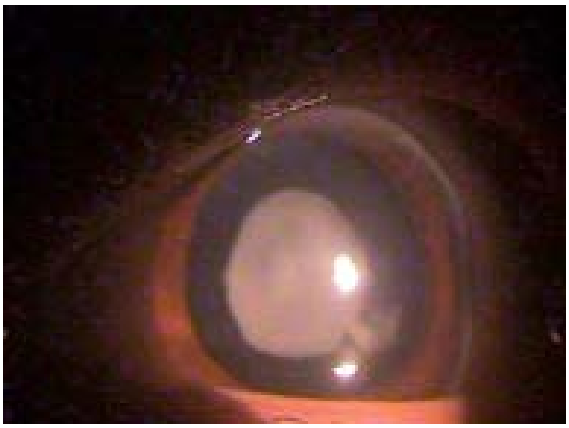


Fig. 16.6: Central nuclear cataract

include a poor development of the dilator pupillae, necrosis of the pigment epithelium of the iris and ciliary body, a chronic low grade granulomatous uveitis, and pigmentary retinopathy. Rubella is capable of producing gross ocular and systemic malformations such as microphthalmos, microcephaly, mental retardation, deafness, and patent ductus arteriosus.

Sutural Cataract (Stellate Cataract)

When opacities involve the sutures of the lens they are termed as *sutural cataract* or *anterior axial embryonic cataract*. The sutural cataract is almost always bilateral, and opacities often have branches and knobs projecting from them. Sutural cataract is inherited in an autosomal dominant pattern.

Coralliform Cataract

Coralliform cataract is also known as *fusiform* or *spindle-shaped cataract*. It is characterized by an anteroposterior spindle-shaped opacity occurring axially with off-shoots resembling a coral (Figs 16.7 and 16.8). It is genetically determined and has a strong familial tendency.

Fig. 16.7: Coralliform cataract

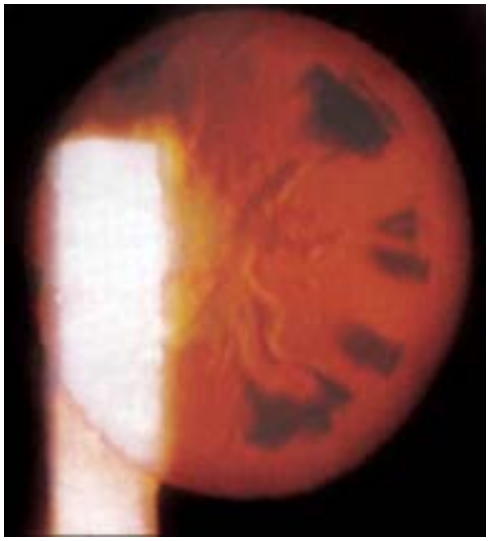


Fig. 16.8: Coralliform cataract on retroillumination
(*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

Zonular Cataract (Lamellar or Perinuclear Cataract)

Zonular cataract is the most common type of cataract accounting for nearly 50% of the total developmental cataracts.The cataract is bilateral and may present at birth or manifest during early infancy or adolescence.

Zonular cataracts, especially the bilateral ones, are usually inherited as an autosomal dominant trait. Congenital nuclear and sutural opacities of lens are often associated with zonular cataract. Maternal metabolic disturbances during pregnancy such as hypoparathyroidism may cause a zonular cataract. Disturbances of calcium metabolism, hypovitaminosis D (rickets) and defective development of enamel of permanent teeth may be associated with the zonular cataract.

Zonular cataract causes variable visual impairment corresponding to the diameter and density of the affected lamella. The diameter of the opacity decreases with time as the nucleus becomes compressed. The opacity affects a

particular lamella so that it encircles the nucleus both anteriorly and posteriorly (Fig. 16.9). When viewed from front, lamellar cataract has a disk-shaped opacity. It is always surrounded by a clear cortex. The opacity is heterogenous and composed of dense and translucent areas, and occasionally made up of small discrete dots. It presents radial projections resembling the spokes of a cartwheel, popularly known as *riders* (Fig. 16.10).

Coronary Cataract

A group of club-shaped opacities in the cortex of the lens is known as *coronary cataract*. The opacities are situated around the equator of the lens encircling the central axis to form a crown or corona (Fig. 16.11). They are non-progressive and vision is seldom affected, except when they are extensive or associated with subcapsular cataract.

Complete or Total Cataract

The complete cataract may be unilateral or bilateral (Fig. 16.12). In complete cataract all the lens fibers are opacified and retina cannot be visualized. Some cataracts are subtotal at birth and progress to become total and cause profound visual impairment.

Membranous Cataract

Membranous cataracts are rare and occur due to absorption of the lens fibers leaving the anterior

Fig. 16.9: Zonular cataract affecting a few lamellae and surrounded by clear cortex

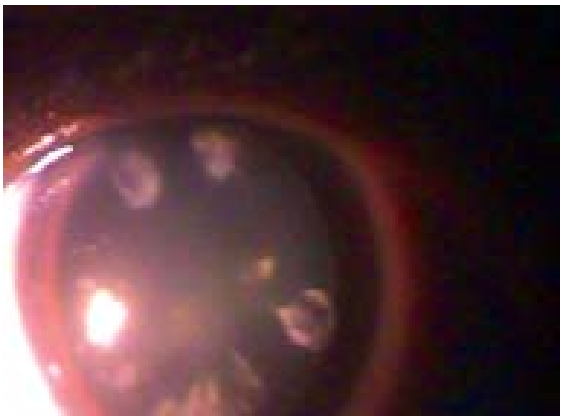


Fig. 16.11: Coronary cataract

Fig. 16.10: Zonular cataract with riders

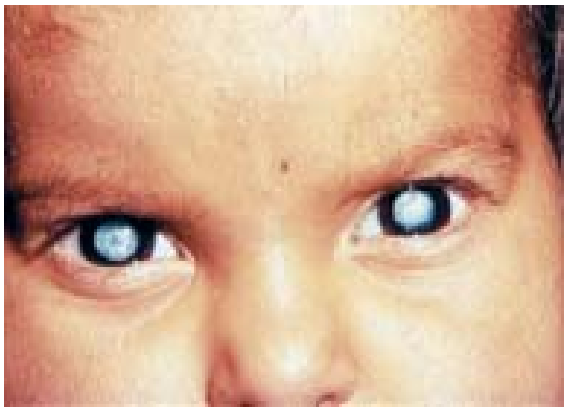


Fig. 16.12: Bilateral total cataract
(*Courtesy*: Dr SK Pandey, Sydney)

and the posterior lens capsule to fuse in a dense membrane leading to marked visual loss.

Treatment of Developmental Cataract

Basic Considerations

The management of developmental cataract requires a wide range of considerations including laterality, extent of opacity and visual impairment, small size of the eyeball, changing axial length of a growing eyeball, lower scleral rigidity, more elastic capsule, high potential of development of amblyopia and frequent late complications of surgery.

1. As most of the developmental cataracts are stationary and do not cause visual impairment, no treatment is required.
2. The use of mydriatics and optical iridectomy are considered obsolete procedures now-a-days.
3. *Bilateral congenital cataracts*: Infants with bilateral congenital cataracts may develop nystagmus at about 3 months of age due to non-development of fixation reflex. Therefore, cataract surgery must be performed on one eye as soon as possible, ideally prior to three months of age, followed by surgery on the fellow eye after 2-4 weeks.
4. *Unilateral congenital cataract*: Infants with unilateral congenital cataract should be operated before 6 weeks of age to prevent deprivation amblyopia. Surgical intervention must be followed by correction of aphakia and amblyopia therapy.
5. *Correction of aphakia: (i) Spectacle phakic correction* can be given after one week of surgery. Aphakic correction and compliance in wearing is often difficult in children younger than one year. However, older children with bilateral aphakia tolerate the spectacles well. *(ii) Contact lenses* are a well-established method of optical correction of monocular or binocular aphakia. Soft hydrophilic contact lenses for extended wear are preferred. *(iii) Intraocular lens implantation* is the most preferred approach in children over 2 years of age. The lens implantation surgery under one year of age is controversial. Posterior chamber lenses are preferred and implanted in the capsular bag.
6. *Prevention of amblyopia*: The risk of amblyopia can be reduced by proper timing of surgery, adequate aphakic correction, postoperative care and management. The parents of the child must be informed and educated that a successful result of developmental cataract surgery largely depends on proper aphakic correction and continued amblyopic therapy.

Prognosis

The visual prognosis of unilateral congenital cataract is less favorable than that of bilateral cataract because unilateral visual deprivation often causes irreversible amblyopia. Complicated congenital cataracts, associated with microphthalmos, persistent hyperplastic primary vitreous or rubella retinopathy, also carry a poor prognosis.

Acquired Cataract

The opacification of already formed lens fibers in the post-natal period is called *acquired cataract*.

Etiology

Acquired cataract occurs due to degeneration of lens fibers following physical or chemical insult. Although the exact etiology is not known, it is suggested that any factor which disturbs the colloid system within the lens fibers or disrupts the intracellular and extracellular water and electrolyte equilibrium can produce cataract.

Epidemiological studies including the Longitudinal Study of Cataract have suggested a number of risk factors for the acquired cataract. They include advanced age, exposure to ultraviolet and infrared radiations, hyperbaric oxygen, diabetes mellitus, dehydration due to diarrhea, and heavy smoking. Deficiency states especially of vitamins E

and C, and carotinoids (antioxidants) expose the lens fibers to free radicals and enhance the effect of ultraviolet light on the formation of cataract.

The role of inheritance in age-related cataract is widely recognized. Genetic mutations in the genes for crystallins and gap-junction proteins cause cataracts in susceptible families. The inheritance is multifactorial, it may show an autosomal dominant, recessive or sex-linked pattern.

Ageing Changes in the Lens

With advancement of age, the lens increases in weight and thickness and the nucleus undergoes hardening (nuclear sclerosis). The proteins of lens fibers (crystallins) aggregate into higher molecular weight proteins. The chemical modification of nuclear lens protein produces a yellow or brown pigmentation. The chemical compositions of ageing lens include decrease concentrations of glutathione and potassium and increase concentrations of sodium and calcium.

Senile Cataract

Age-related cataract is by far the most common variety occurring bilaterally often asymmetrically in persons above sixty years of age. Both males and females are equally affected. Senile cataract is familial and shows a strong hereditary tendency manifesting at an earlier age in successive generations.

Types

Senile cataract occurs in three forms:
1. Cortical
2. Posterior subcapsular, and
3. Nuclear.

Clinical Features

In the initial stage of cataract almost all patients remain symptom-free. Common symptoms of cataract include glare, black spot before eyes, distortion of objects, polyopia, colored halos, and a variable degree of visual impairment.

Glare or dazzling is common under bright light conditions. The patient feels handicapped in night driving because the posterior subcapsular opacities obscure the pupillary aperture when miosis is induced by the bright light.

Black spots may be perceived by the patient due to the presence of lenticular opacities.

Distortion of objects occurs in the early stages of cataract formation due to changes in the refractive indices of the lens fibers causing irregular refraction.

Polyopia occurs due to an irregular refraction and the patients often complain of seeing many moons in the sky or perceive many images of an object. Colored halos may be seen by some patients due to the hydration of lens fibers.

Impairment of vision is variable depending on the site, extent and progress of the lens opacity. Early cortical cataract does not cause any impairment of vision while mature cortical cataract and posterior polar cataract lead to marked visual loss. The patients with immature cortical cataract can see better in day light but feel handicapped in twilight owing to pupillary dilatation. Conversely, the patients with nuclear cataract have better vision in dimlight.

Second sight develops in patients of nuclear sclerosis. Difficulty in near work occurs in old age due to loss of accommodation. In patients with lenticular sclerosis index myopia develops leading to deterioration of distant vision but the patient starts seeing better for the near and may even give up the use of presbyopic glasses, a phenomenon known as *second sight*.

Senile Cortical Cataract

Pathogenesis

It is postulated that the senile cortical cataract results from altered physiochemical processes

within the cortex of the lens. The main processes involved in cataract formation are hydration, and replacement of soluble by insoluble proteins. Fine droplets of fluid and water cleft can be seen under the capsule on the slit-lamp in the initial stages of cataractogenesis. The initial process is reversible to some extent, but later the lens swells up and becomes opaque (intumescent). At this stage denaturation of protein of the lens fibers occurs, altering them chemically from non-coagulable to readily coagulable form. Thus, dense and irreversible opacities are produced. Histologically, cortical cataracts present with hydropic swelling of the lens fibers and accumulation of eosinophilic material between them.

The clinical course of the development of senile cataract can be divided into 5 stages.

1. *Stage of lamellar separation* or *presenile change* is characterized by the collection of fluid between the lens fibers resulting in lamellar separation which can be demonstrated on slit-lamp biomicroscopy. The hydration causes change in the refractive index of the cortex. Generally, there are no symptoms except the patient becomes slightly hypermetropic.
2. *Incipient stage* presents with white radial or wedge-shaped spokes of opacities in the periphery of the lens (Fig.16.13). Such opacities are also known as *cuneiform cataract*. They are commonly seen in the lower nasal quadrant. If the opacities are too peripheral their recognition in undilated pupil becomes difficult, but later their apices extend beyond the normal pupillary margin. The incipient cortical cataract changes the refractive indices of the lens fibers causing irregular refraction, hence, polyopia (many images of an object), colored halos, and visual disturbances are common in this stage of cataract formation. The patient has a defective vision especially in the evening or night owing to the dilatation of pupil. The opacities appear gray on oblique illumination and black against the red glow of the fundus when seen by an ophthalmoscope or on plane mirror examination.

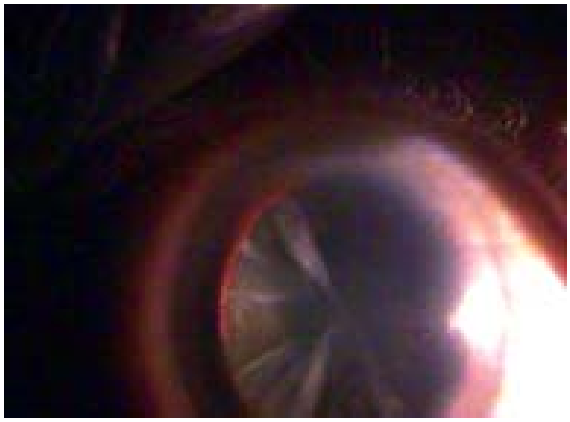


Fig. 16.13: Wedge-shaped spokes of opacities in cortex

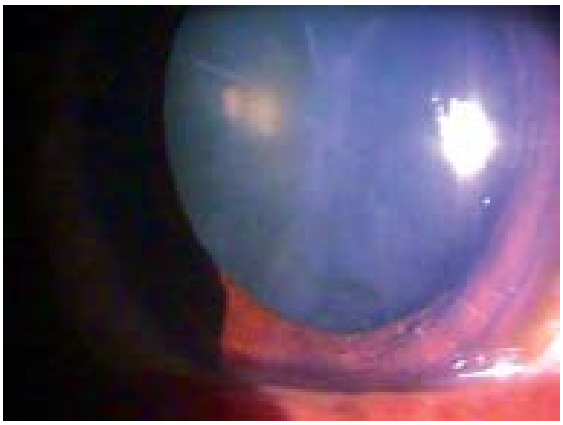


Fig. 16.14: Intumescent cataract

3. *Intumescent stage* is characterized by diffuse and irregular lenticular opacities due to the hydration of deeper cortical layers. The progressive hydration causes swelling and opacification of the lens which is called as *intumescent cataract* (Fig. 16.14). Such a lens can push the iris forwards in an already existing shallow anterior chamber and may produce embarrassment of the chamber angle leading to secondary angle-closure glaucoma.

   Upto this stage the lens is not completely opaque. There remains a clear zone of lens fibers between the pupillary margin of the iris and the lens opacity (immature cataract). If a

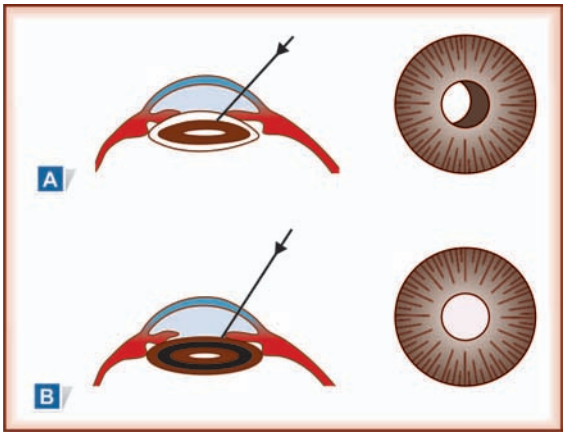


Fig. 16.15: (A) Immature cataract with iris shadow, (B) Mature cataract without iris shadow

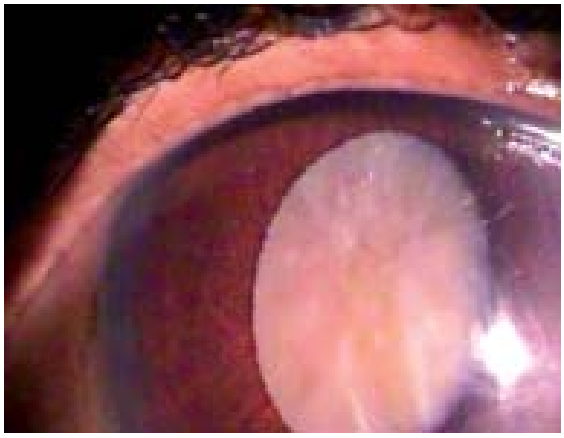


Fig. 16.16: Mature cataract

beam of light is thrown upon the eye obliquely, the iris casts a shadow upon the gray opacity. When the lens is completely opaque, the pupillary margin lies almost in contact with the opaque lens and the iris does not cast any shadow. The presence and the absence of the iris shadow (Fig. 16.15) is a useful sign in differentiating immature and mature cataracts, respectively (Table 16.1).

Table 16.1: Differentiation between immature and mature cataract

Features	*Immature*	*Mature*
1. Vision	CF or above	Usually HM
2. Iris shadow	Present	Absent
3. Purkinje's fourth image	Present	Absent

CF: Counting fingers
HM: Hand movements

4. *Mature cataract* presents with opacification of the entire cortex and the lens becomes totally opaque. Such a cataract is known as *mature* or *ripe* (Fig. 16.16). The visual acuity is grossly reduced to hand movements or light perception.

   Sometimes polychromatic crystal-like appearance of the cataractous lens is seen as a normal ageing phenomenon. It is termed as *Christmas tree cataract*. This type of cataract is commonly seen in dystrophia myotonica and tetany.

   The rate of development of senile mature cataract varies from individual to individual. It is not uncommon to witness a very rapid development and maturation of cataract in young individuals suffering from diabetes mellitus or iridocyclitis, while in others immature cataract may remain stationary for many years and never reach maturity. The progression of cataract should be recorded on periodical examination either by slit-lamp photography or by careful drawings.

5. *Hypermature cataract* is usually of 2 types:
   a. *Morgagnian type:* When a mature cataractous lens is not extracted from the eye, the stage of hypermaturity sets in. The soft cortex liquifies and the hard nucleus sinks to the bottom (Fig. 16.17). The pultaceous cortex appears milky and the nucleus looks as a brown mass. The nucleus changes its position with the movements of the head. Such a cataract is known as *Morgagnian cataract*. Occasionally, uveitis may develop in patients with a hypermature cataract.
   b. *Sclerotic type:* Sometimes the loss of fluid from the mature cataractous lens continues and

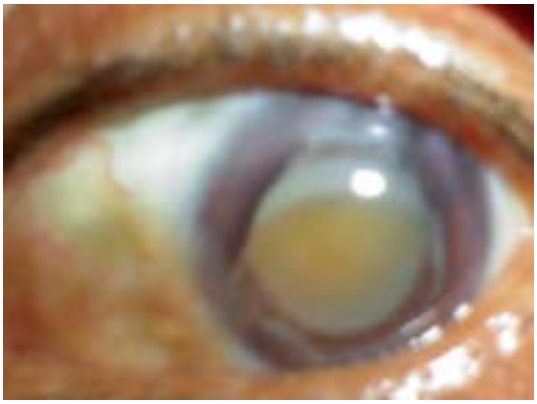


Fig.16.17: Morgagnian cataract (*Courtesy*: Prof. Manoj Shukla and Dr Prashant Shukla, AMUIO, Aligarh)

the lens becomes very much inspissated and shrunken. The lens appears yellow owing to the deposition of cholesterol crystals, while dense capsular cataract is formed at the anterior pole due to the vicarious proliferation of the anterior epithelial cells. The shrinkage of the lens leads to the deepening of the anterior chamber and tremulousness of the iris. The associated degeneration of the zonule may lead to dislocation of the lens.

Posterior Subcapsular Cataract

Posterior subcapsular or cupuliform cataracts develop in the posterior cortical layer and are often axial. The cataract manifests at a younger age than cortical or nuclear cataract.Owing to axial location, the patients with posterior subcapsular cataract complain of glare and poor vision in bright light. In early stage, slit-lamp examination through a dilated pupil reveals a subtle iridescent sheen in the posterior cortical layers (Fig. 16.18). Later granular opacities or a plaque-like opacity may develop which can be confirmed on retro-illumination (Fig. 16.19). Histopathology indicates that swollen lens epithelial cells have migrated to the posterior subcapsular area.

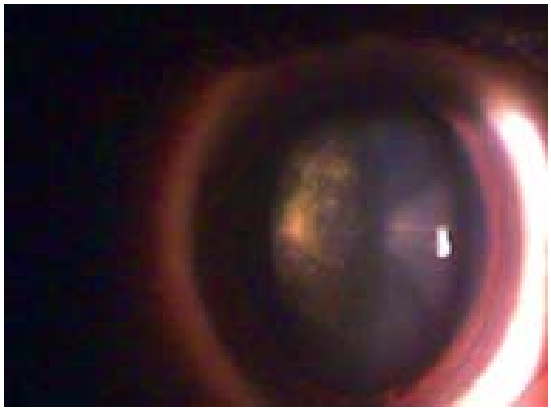


Fig. 16.18: Posterior subcapsular cataract

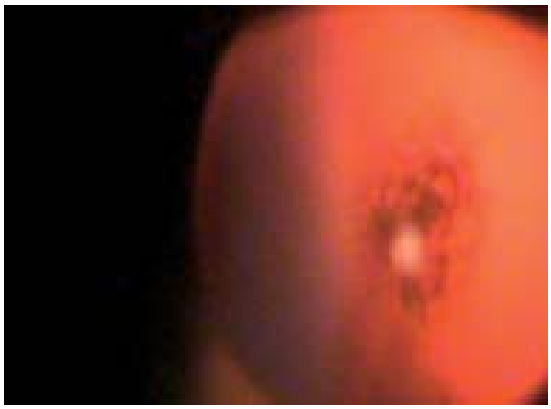
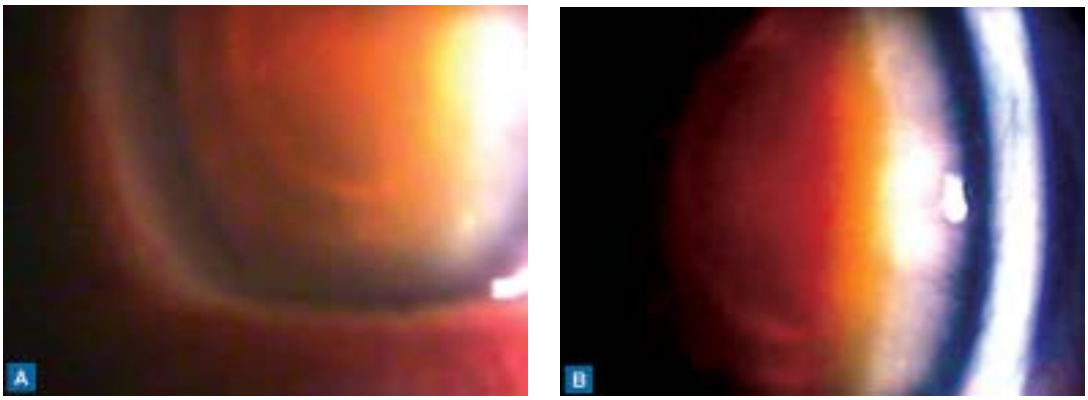


Fig. 16.19: Posterior subcapsular cataract on retroillumination

Senile Nuclear Cataract

Senile nuclear cataracts are bilateral and asymmetrical and develop as a result of slow but progressive sclerosis—an accentuation of physiological phenomenon. The changes are marked in the nucleus of the lens which becomes hard and yellow as the age advances (Figs 16.20A and B). The sclerosed fibers of the nucleus may become brown (*cataracta brunescence*) or black *(cataracta nigra)* owing to the deposition of melanin derived from tryptophan group of amino acids. The nuclear sclerosis increases the refractive index of the lens and the patient becomes myopic causing impairment of distant vision. Initially the nucleus



Figs 16.20A and B: Nuclear cataract (Cataracta brunescence)

appears more refractile and translucent, gradually the central part of the lens becomes completely opaque and later the opacity may involve the peripheral cortex too. A mature nuclear cataract may extend almost to the capsule and the entire lens appears as nucleus. Ophthalmoscopy may not reveal any fundus glow. In cataracta brunescence or black cataract the pupillary reflex appears black. However, most nuclear cataracts take long time to reach maturity and they seldom become hypermature. Nuclear cataracts show homogeneity of lens nucleus with loss of cellular laminations on histopathological examination.

The distinguishing features between senile immature nuclear cataract and immature cortical cataract are summarized in Table 16.2.

Complicated Cataract

The complicated cataract is characterized by an ill-defined opacification of the posterior cortical area giving a bread-crumb appearance.

Etiology

The complicated cataract is secondary to the inflammatory or degenerative diseases of the eye such as iridocyclitis, choroiditis, high myopia, retinal detachment and primary pigmentary degeneration of the retina. A small anterior subcapsular lens opacity, *glaucomflecken*, may develop as a result of ischemia in patients with acute congestive glaucoma.

Table 16.2: Differentiation between senile immature nuclear cataract and immature cortical cataract

Features	*Senile immature nuclear cataract*	*Senile immature cortical cataract*
1. Vision	Poor in day time, may improve with minus lenses	Poor in night time, may improve with plus lenses
2. Age of onset	Usually starts in forties	Usually starts in fifties
3. Lens opacities		
a. Site	Central	Peripheral
b. Shape	Nearly round or oval	Wedge-shaped spokes
c. Color	Brown or black	Usually white
d. Maturity	Takes long time to become mature	Matures relatively early
e. Hypermaturity	Rarely becomes hypermature	If unattended, reaches hypermaturity

Clinical Features

The vision is markedly impaired since the opacity is situated near the nodal point of the eye. The cataract usually starts near the posterior pole, and the posterior cortical region shows an opacity with irregular margins extending towards the equator and the nucleus (Fig. 16.21). The slit-lamp examination exhibits a characteristic rainbow display of colors called *polychromatic luster*—a diagnostic sign of complicated cataract. The complicated cataract may remain stationary or may progress peripherally affecting almost all the posterior cortex and may extend axially to involve the entire lens (Fig. 16.22). A mature complicated cataract is often cortical in form but later signs of hypermaturity supervene.

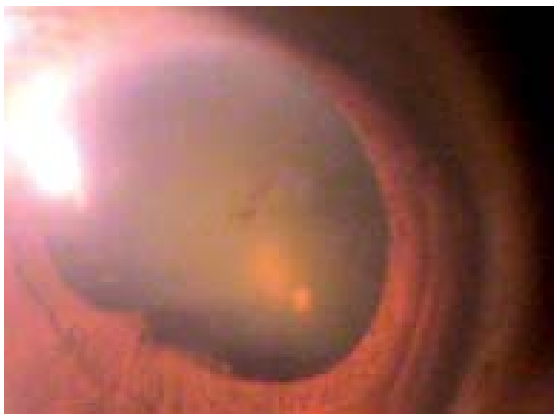


Fig. 16.21: Complicated cataract caused by uveitis

Traumatic Cataract

Mechanical injury, radiation, electrical current and chemical agents are capable of causing traumatic lens changes. Besides damaging the ocular surface, chemical injuries can cause cataract. A cortical cataract may develop as a result of delayed effect of alkali burn. The mechanical injury may be either a contusion or a penetrating injury.

Contusion or Blunt Injury

A blunt injury to the eye may occur by fist or a tennis or a cricket ball and may produce Vossius ring, traumatic cataract and subluxation or dislocation of lens.

Vossius ring: Contusion leads to the release of pigments from the pupillary ruff that get imprinted onto the anterior surface of the lens in a ring form, called *Vossius ring*. The size of the ring is nearly same as the diameter of the constricted pupil. The presence of the ring is suggestive of a blunt trauma.

Traumatic cataract: A blunt trauma may cause traumatic cataract either as an early or as a late sequela. Typically a concussion cataract is rosette- or stellate-shaped (Fig. 16.23) and axial in location

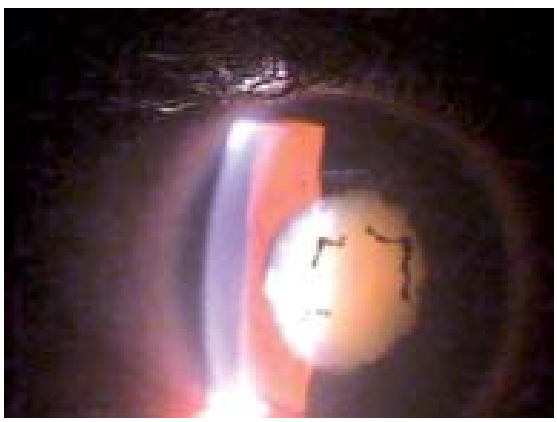


Fig. 16.22: A total complicated cataract with broken posterior synechiae

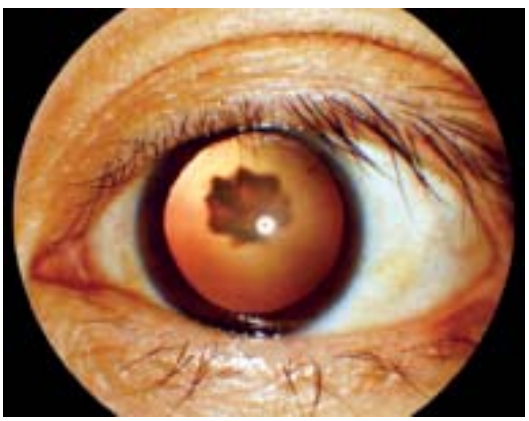


Fig. 16.23: Stellate-shaped cataract following blunt trauma (*Courtesy*: Mr S. Kanagami, Tokyo)

involving the posterior lens capsule. The cataract may progress to opacification of the entire lens.

Subluxation and dislocation of lens: Sudden expansion of globe in an equatorial plane may occur during blunt trauma causing disinsertion of zonular fibers resulting in subluxation (Fig. 16.24) or dislocation of the lens.

Penetrating injury

Penetrating injury may occur with a neddle, thorn, arrow or a flying foreign body. The patient often complains of sudden blurring of vision and eye becomes red and soft.

A small perforating injury of the lens capsule may heal and only a small localized opacity is formed. However, more frequently, a penetrating injury results in swelling of the lens and the entire lens becomes cataractous.

When a small non-metallic foreign body perforates the cornea and the lens capsule and gets lodged within the lens, intralenticular foreign body, it may cause a focal cortical cataract without much reaction. The intraocular metallic foreign bodies composed of iron or copper often cause cataract and ocular tissue discoloration.

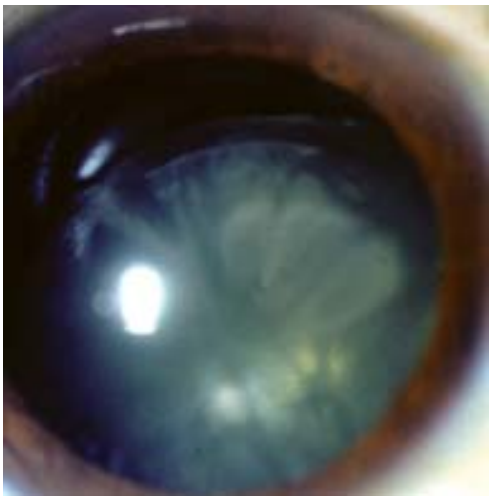


Fig.16.24: Traumatic anterior subluxation of lens

A retained intraocular iron foreign body can lead to *siderosis bubli*, a condition marked by deposition of iron in the lens epithelium, iris, trabeculum and retina. The lens epithelium and fibers show a rusty brown discoloration. Later cortical cataract and retinal dysfunction may develop.

A retained intraocular copper foreign body results in *chalcosis*, a condition characterized by sunflower cataract formation associated with yellow or brown pigmentation of Descemet's membrane and the anterior lens capsule. The foreign body composed of pure copper can induce a severe inflammatory reaction in the eye.

Radiation Induced Cataract

1. *Ionising radiation*: Actively growing lens cells are extremely sensitive to ionizing radiation. An early radiation induced cataract is characterized by punctate opacities within the posterior capsule and feathery anterior subcapsular opacities radiating towards the lens equator.
2. *Infrared radiation*: The intense heat of infrared radiation may cause true exfoliation of the anterior lens capsule and cataract formation, also known as *glass blower's cataract*.
3. *Ultraviolet radiation*: Ultraviolet radiation in the range of 290-320 nm is considered as a risk factor for the development of cortical and posterior subcapsular cataracts.
4. *Microwave radiation*: There is an inconclusive evidence to suggest that the microwave radiation causes cataract in human beings.

Electrical Injury

A powerful electrical shock can cause coagulation of lens proteins and cataract formation. Vacuoles and linear opacities develop in the anterior subcapsular cortex. The cataract may remain stationary or progress to maturation.

Metabolic Cataract

The metabolic cataract is caused by endocrine disorders and biochemical abnormalities. Some are associated with inborn errors of metabolism such as galactosemia.

Diabetic Cataract

Diabetes mellitus can affect the clarity of lens, its refractive index and accommodation. Diabetic cataracts can be of two types*: true diabetic cataract* (snowflake cataract) and *senescent cataract.*

In diabetic patients, significant amount of sorbitol and fructose get accumulated in the lens which in turn increase the intralenticular osmolarity and draw water into the lens.

True diabetic cataract occurs due to the osmotic overhydration of the lens. It is bilateral and seen in uncontrolled juvenile diabetics. In the early stage, a number of fluid vacuoles appear under the capsule, but later snowflake opacities develop all over the cortex giving a milky-white appearance to the lens. The control of diabetes may partially resolve the opacities, but more often they become confluent to make the entire lens opaque.

Senescent cataract occurs earlier, rapidly and more frequently in diabetics than in non-diabetics.

Tetanic Cataract

Inadvertent removal of parathyroid gland during thyroidectomy leads to the deficiency of parathyroid hormone and calcium (idiopathic hypocalcemia) resulting in cataract formation. The cataract is marked by the appearance of small discrete opacities in the cortex which are separated from the capsule of the lens by a clear zone. These opacities coalesce to form large shining crystalline flakes.

Galactosemic Cataract

Galactosemia is an inborn error of carbohydrate metabolism characterized by inability to metabolize galactose due to the deficiency of the enzyme galactose-1-phosphate uridyl transferase. Galactose is reduced to dulcitol within the lens, the accumulation of which causes lens opacities. The dust-like lenticular opacities manifest soon after birth (within 2 months) and the nucleus and deep cortex become opaque causing a classical "oil droplet" appearance on retrolumination. Besides bilateral cataract, other clinical manifestations include mental retardation, splenohepatomegaly, jaundice and ascites. Opacities are initially lamellar, but eventually become total. However, the lens changes are reversible. Dietary exclusion of milk and food containing galactose and lactose during first 3 years of life usually prevents galactosemic cataract.

Lowe's Syndrome

Lowe's syndrome is an inborn error of amino acid metabolism predominantly affecting the male children. The syndrome is characterized by mental retardation, renal dwarfism, muscular hypotonia, osteomalacia, congenital cataract and glaucoma. Congenital cataract is almost always found. The lens opacities may be nuclear, lamellar or total.

Wilson's Disease

Wilson's disease is an autosomal recessive disorder of copper metabolism caused by the deficiency of alpha-2 globulin, ceruloplasmin, resulting in a widespread deposition of copper in the body. It is characterized by hepatosplenomegaly, tremors, spasticity, mental changes, Kayser-Fleischer (KF) ring and cataract. The cataract is not as frequent as KF ring. A characteristic lenticular opacity may develop in the anterior capsular region. It has a sunflower pattern and is bright red-brown in color. It represents deposition of metallic copper in the lens capsule. Treatment with D-penicillamine may remove the copper.

Dermatogenic Cataract

Dermatogenic cataracts are bilateral and occur in young age.

Atopic Dermatitis

Atopic dermatitis is a chronic erythematous skin disorder associated with increased level of IgE. Cataract may develop in upto 25% patients with atopic dermatitis. The atopic cataracts are bilateral and develop in the third decade of life. The opacities are anterior subcapsular involving the pupillary area and resemble a shield-like plaque that may gradually involve the entire lens.

Rothmund's Syndrome

Rothmund's syndrome is an uncommon recessively inherited skin disease predominantly affecting the females. The disease is characterized by eruptive and exudative skin lesions beginning in the first year of life, bony defects, sparse hair growth, hypogonadism and bilateral zonular cataract.

Werner's Syndrome

Premature senility, diabetes insipidus, dwarfism, endocrine disturbances and bilateral posterior subcapsular cataract characterize Werner's syndrome. The disease has a recessive inheritance and a high incidence of malignancies.

Cataract Associated with Maternal Infections

Congenital cataract may be due to maternal infections like rubella, toxoplasmosis and cytomegalic inclusion disease.

Rubella

Rubella infection of non-immune mothers, in the first trimester of pregnancy, causes malformation in approximately 50% of fetuses. The fetal infection is probably a result of maternal viremia. Spontaneous abortion and still-birth are common. The classical manifestations of rubella infection are congenital heart defects, deafness and cataract (rubella triad). The cataract may be unilateral or bilateral and usually present at birth. The rubella cataract is characterized by a pearly-white nuclear opacification. Sometimes it is more diffuse causing a total opacification of the lens. Other ocular features of rubella include diffuse pigmentary retinopathy, glaucoma, microphthalmos and corneal clouding.

Drug Induced Cataract (Toxic Cataract)

Long-term administration of corticosteroids, phenothiazine and miotics may induce lens changes.

Corticosteroid Cataract

Posterior subcapsular opacities may occur following topical, subconjunctival and systemic long-term use of corticosteroids. The clinical features of posterior subcortical cataract are indistinguishable from posterior subcapsular form of senile cataract.

Phenothiazine Cataract

The administration of chlorpromazine and thoridazine can cause axial pigmented opacities in the anterior lens epithelium.

Miotic Cataract

It has been reported that nearly 20% patients after 55 months of pilocarpine therapy may develop cataract. Vacuoles may appear in the anterior lens capsule and epithelium which may progress to posterior cortical or nuclear cataract.

Cataract Associated with Systemic Diseases

Dystrophia Myotonica

Fine dust-like opacities interspersed with polychromatic iridescent spots develop in the lens cortex underneath the capsule in 90 percent of the patients with dystrophia myotonica. They may or may not be associated with posterior subcapsular stellate (Christmas tree) opacities. The systemic features of dystrophia myotonica include hypercontractility and difficulty in relaxation of skeletal muscles, weakness of facial muscles and cardiac lesions.

Down's Syndrome

Multiple punctate and flake-like opacities are found early in life in the cortex of the lens in Down's syndrome. Small percentage of cases present with congenital cataract involving the fetal nucleus. Lens sutures often appear more prominent and gray. Brushfield spots, white or gray thickening of the mid-peripheral iris arranged in a ring, concentric with the pupil, are found in 85 percent patients of Down's syndrome.

Evaluation and Treatment of Cataract

Each case of cataract must be thoroughly evaluated for the extent of visual impairment and its effect on day-to-day working of the patient. It should be ascertained that whether the lens opacity corresponds to the degree of visual loss. The accompanying eye disease, especially glaucoma, may cause a disproportionate visual impairment. A number of factors such as type of cataract, size of the pupil and degree of myopia often affect the visual acuity of the patient.

Management

Prophylaxis

Smoking, exposure to ultraviolet light, and diabetes, are known risk factors for the development of cataract. Cigarette smoking is linked to an increased risk for nuclear sclerosis. Cortical cataract is common in people who work in open farms and fields and are exposed to ultraviolet light. Diabetics are at a higher risk for both cortical and posterior subcapsular cataracts.

Preventive strategies include avoiding smoking, abstinence from alcohol, judicious use of steroids, use of protective glasses to shield against sunlight, and strict glycemic control in diabetic patients. The role of asprin and antioxidants in delaying the process of cataractogenesis remains inconclusive.

Medical Management

In spite of the fact that there is no medical treatment for cataract, a number of agents such as N-acetyl carnosine (NAC), potassium iodide drops, cineraria maritima, etc. are emperically and enthusiastically being recommended by the practitioners. Adoptation of following simple measures can temporarily improve the vision in patients with immature cataract.

1. *Glasses*: Prescription of suitable glasses after refraction may improve both the near and the distant vision. Patients with nuclear cataract are benefited with the use of tinted glasses or sun goggles.
2. *Illumination*: The visual efficiency of a patient with nuclear cataract improves by keeping the light behind the patient. In cortical cataract, small pupil cuts off the lens opacities, therefore, a bright source of light is kept in front while working.
3. *Mydriatics*: Besides the arrangement of illumination, instillation of a weak mydriatic such as cyclopentolate 0.5% , phenylephrine 2.5% or tropicamide 1%, may improve the visual function in patients with small axial cataracts by allowing the light to pass through the peripheral portion of the lens, although it carries the risk of precipitation of acute angle-closure glaucoma in patients with an occludable angle of the anterior chamber.

Pupillary dilatation can also be achieved by laser pupilloplasty.

4. *Low vision aids*: A limited visual improvement can be obtained by providing optical visual aids. Handheld magnifiers, spectacle-mounted telescopes and high plus spectacles may be used for reading or close work.

Once the lens opacification has occurred, no amount of treatment, local or general, will reverse it. It may be re-emphasized that presently there is no medical treatment for cataract. The only effective treatment of cataract is its operative removal. The details of cataract operation are described in the Chapter on *Operations Upon the Eyeball and its Adnexa*.

APHAKIA

The absence of the lens from its normal position is called *aphakia*.

Etiology

Surgical removal of the lens is by far the commonest cause of aphakia. The lens is dislocated by a sharp needle in the vitreous cavity in *couching*. Spontaneous absorption of the lens in children is observed following penetrating injury or after performing needling for congenital cataract. Dislocation of the lens causes aphakia. It may occur due to blunt injury or as a result of degeneration of the zonule as found in longstanding cases of anterior uveitis and high myopia. Rarely, the lens may be absent congenitally.

Clinical Features

Deep anterior chamber, tremulousness of the iris (iridodonesis) and a jet-black pupil (after intracapsular cataract extraction) are the classical signs of aphakia. A scar mark at the limbus or in the peripheral cornea and a coloboma of the iris (if surgical iridectomy is done) are found in surgical aphakia. The absence of the 3rd and the 4th Purkinje's images can also be noticed.

The removal of the lens causes complete loss of accommodation, and the eye becomes extremely hypermetropic resulting in deterioration of vision both for near and distance. The refractive power of the eye becomes about + 44 D (the phakic eye power is about + 60 D). Besides hypermetropia, some acquired astigmatism (against-the-rule) occurs owing to the scarring of the superior incision.

Normally, the lens absorbs the near ultraviolet light (300-400 nm) which is transmitted by the cornea. Occasionally, aphakic patients develop the sensation of transient red vision following exposure to the ultraviolet light; such a vision is known as *erythropsia* wherein objects appear red, but visual acuity is seldom affected.

Treatment

Spectacle correction: The refractive error in aphakic eye is determined by retinoscopy and glasses are prescribed on the subjective acceptance. If preoperatively the eye was emmetropic, a power of + 10 D is generally required to correct the acquired hypermetropia, while in ametropic eye the previous error of refraction will modify the correction (add the hypermetropic error and subtract the myopic). The astigmatic error (against-the-rule) usually amounts to 1 to 3 D cylinder and is corrected by prescribing the cylinder at 180° axis (in case of a superior limbal or corneal incision). The spectacle lens causes enlargement of the retinal image of an object by 25 to 30% more than that of the normal eye. Hence, diplopia is common in uniocular aphakia with good vision in the fellow eye.

The aphakic patients have to adapt to the optical problems of aphakic spectacles. Besides enlargement of the image, the problems include ring scotoma with jack-in-the-box phenomenon and spherical aberration with pin-cushion distortion. These problems result in poor hand-

to-eye coordination and spatial disorientation. A hyperaspherical aphakic lens has been developed which increases the field, reduces the magnification and moves the ring scotoma more peripherally.

Contact lenses: Many problematic aphakic cases can be fitted with contact lenses. Contact lenses give better coordination and mobility, because the magnification is only 7 to 9% and there is no spherical aberration and jack-in-the-box phenomenon. However, they require dexterity to insert and good hygiene to maintain. Correction with contact lenses in patients with monocular aphakia restores some degree of binocular vision.

Intraocular lens implantation: Aniseikonia (difference between the size of images formed in two eyes) can further be reduced to 1 to 2% by intraocular lens (IOL) implantation either during cataract surgery (primary IOL implantation) or sometimes after the surgery (secondary IOL implantation). The former is preferred to avoid second operation and its inherent complications. The intraocular lenses are almost free from the disadvantages of aphakic spectacles and contact lenses. There is minimal magnification of the image and practically no optical aberration. The IOLs offer a full field of vision and are free from the hassle of daily insertion and removal. In view of these definitive advantages in terms of vision and convenience over other available methods of aphakic correction, increasing number of ophthalmic surgeons are combining cataract extraction with IOL implantation.

As there is a complete loss of accommodation in aphakia, +3 or +3.5 D spherical lenses are added on the distance correction for the near work. Hence, the patient requires an additional pair of glasses for reading. Recently, bifocal contact lenses and bifocal intraocular lenses are being made in different designs to provide aphakic corrections.

AFTER CATARACT OR SECONDARY CATARACT

After cataract is an opaque membrane in the pupillary area formed by the remnants of the lens cortex and capsule (Fig. 16.25) following extracapsular lens extraction. With the development of postoperative iridocyclitis or hyphema in some cases, organized exudates and fibrin are added to the membrane. The anterior lens epithelium makes an abortive attempt to form the lens fibers which are often opaque. Sometimes, the cubical cells lying between the anterior and posterior layers of the capsule form a dense ring known as the *ring of Sommerring*, while at other occasions the cells become markedly hypertrophic and appear like balloons which are known as *Elschnig's pearls*. A thin after cataract seldom interferes with the vision. However, the presence of a dense after cataract in the pupillary area causes considerable visual impairment. Occasionally, the membrane may cause pupillary block glaucoma.

Management

The development of after or secondary cataract is preventable. The surgeon must try to remove all

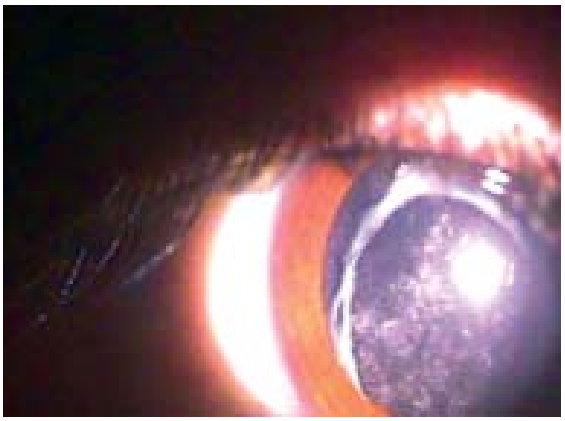


Fig. 16.25: After cataract

the lens material meticulously during the surgery. Postoperatively, use of topical steroid and cycloplegic is necessary to prevent anterior uveitis.

After cataract cases need surgical intervention. Thin after cataract can be incised with a Ziegler's knife or a microvitreoretinal (MVR) blade. The incision in the membrane should be given perpendicular to tension lines so that the margins gape widely. Thick after cataract in chidren requires pars plicata membranectomy and vitrectomy. Presently Neodymium: Yttrium-Aluminum-Garnet (Nd: YAG) laser is used to make a non-invasive opening in the opacified posterior lens capsule.

DISLOCATION OR SUBLUXATION OF LENS

The dislocation or luxation of lens may be partial (subluxation) or complete (dislocation).

Etiology

The etiology of subluxation of the lens is variable. Congenital subluxation of the lens (ectopia lentis) occurs in Marfan's syndrome, Weill-Marchesani syndrome, homocystinuria and Ehlers-Danlos syndrome. Trauma, high myopia, chronic iridocyclitis, buphthalmos and hypermature cataract are other causes of subluxation of the lens (Table 16.3).

Table 16.3: Causes and sites of subluxated lenses

Cause	*Site of displacement*
Ectopia lentis	Complete or partial displacement of lens into the anterior chamber
Marfan's syndrome	Lens is displaced often superotemporally but the zonule is intact
Weill-Marchesani syndrome	Lens is round and small and displaced inferiorly
Homocystinuria	Zonule is torn and lens is displaced often inferonasally
Trauma	Zonule is torn partially or completely causing subluxation or dislocation of lens
Pseudoexfoliation	Zonule is weak due to loose insertion on the lens and the lens may be displaced
Buphthalmos	Enlargement of the eye causes stretching of the zonule and displacement of lens
High axial myopia	Zonular defect may occur due to excessive elongation of the eye causing lens displacement

Clinical Features

The *subluxation of the lens* occurs when fibers of the suspensory ligament or the zonule are torn in one segment. Although the lens is displaced opposite to the segment wherein the fibers are torn, it still remains in the pupillary area. A subluxated lens may remain transparent or become opaque (Fig. 16.26) and cause defective vision and uniocular diplopia. An irregular depth of the anterior chamber, tremulousness of the iris and the lens, and presence of both phakic and aphakic pupillary areas (which causes diplopia) are

Fig. 16.26: Medially subluxated opaque lens

diagnostic. The lens may be subluxated upwards (Figs 16.27A and B) or downwards (Fig. 16.28). The edge of the lens in the pupillary area appears as a black crescent on ophthalmoscopy.

When all the fibers of the zonule are torn, the lens may float into the anterior chamber or drop in the vitreous (*luxation* or *dislocation of the lens*). Subconjunctival dislocation of the lens may occur

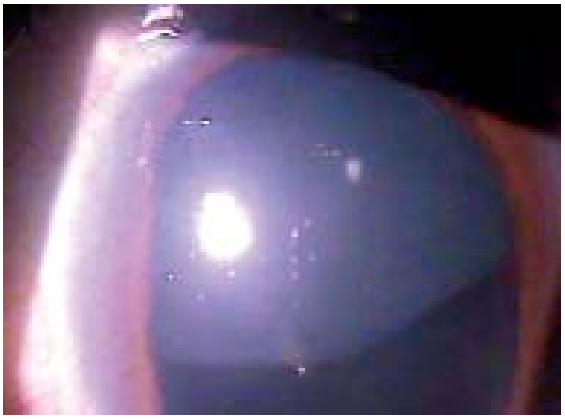


Fig. 16.27A: Superiorly subluxated lens

Fig. 16.27B: Superiorly subluxated lens on retroillumination

Fig. 16.28: Downward subluxation of the lens

following traumatic rupture of the sclera. A clear dislocated lens in the anterior chamber appears like a drop of oil. It interferes with the drainage of the aqueous humor and causes secondary glaucoma *(phacotopic glaucoma)*. The posterior dislocation of the lens in the vitreous cavity results in aphakia. The lens can be located in the vitreous with the help of an indirect ophthalmoscope. The posteriorly dislocated lens may induce iridocyclitis due to irritation of the ciliary body.

Management

The management of dislocation or subluxation of the lens requires meticulous evaluation of an individual case. When the subluxated lens is clear and does not cause any symptom, spectacle correction is advised; an aphakic correction gives better visual acuity than the phakic.

The dislocated lens in the anterior chamber should be removed as early as possible to prevent the peripheral anterior synechia formation. No attempt should be made to fish out the lens from the vitreous cavity as it ends up in gross vitreous loss, retinal break formation and subsequent retinal detachment. In the posterior dislocation of the lens, the accompanied iridocyclitis is treated with cycloplegic, and steroids. The lens from the vitreous cavity is removed by pars plana or limbal route. An anterior chamber IOL or a scleral fixated posterior chamber IOL is implanted. The subluxated cataractous lens can be dealt with pars plana lensectomy or cataract extraction using a capsule tension ring (CTR) and implantation of an IOL with good visual prognosis.

CONGENITAL ANOMALIES OF THE LENS

Besides congenital cataracts and ectopia lentis, lenticonus, coloboma of the lens and microspherophakia may occur.

Lenticonus is a rare anomaly in which posterior or anterior pole of the lens assumes a conical shape, the former is more common. The change in the curvature of the lens results in myopia. The condition is best diagnosed on slit-lamp biomicroscopy.

Coloboma of the lens is characterized by a notch-shaped defect (Fig. 16.29) usually at the inferior margin. It is due to a localized aberration in the development of the suspensory ligament of lens.

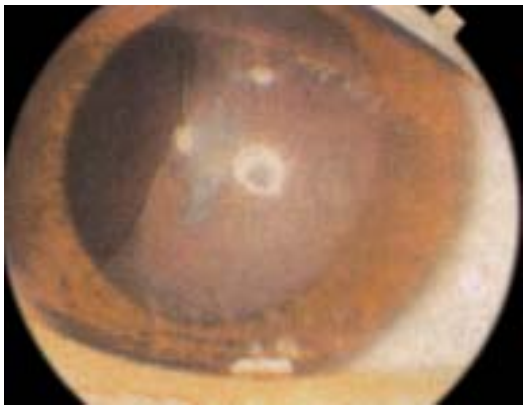


Fig. 16.29: Coloboma of lens

Microspherophakia is a condition in which the lens is small and spherical. It is a developmental anomaly wherein the zonular attachment is lacking. It is recessive in trait and found in Weill-Marchesani syndrome. The small lens can cause a pupillary block particularly with inadvertent use of miotics (inverse glaucoma).

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CHAPTER

17 Diseases of the Vitreous

ANATOMY

Vitreous occupies the posterior segment of the eyeball. It is attached to the edges of optic disk, macula and a zone near the ora serrata. Anteriorly, it is firmly adherent to the posterior surface of the lens in adolescents but later a capillary space develops between the two (Berger's space).

Vitreous is a transparent, colorless, gelatinous mass. It contains water (99%) and interfibrillar material (hyaluronic acid). The structural framework within the vitreous provides it considerable tensile strength and elasticity for maintaining its form. The interaction between hyaluronic acid and the collagen fibrils is responsible for the gel form of the vitreous body.

The vitreous body has three parts: the *main mass* of the vitreous, the *base* of the vitreous (attachment to ora serrata) and the *hyaloidean vitreous* (Fig. 17.1). The vitreous contains a few fusiform cells known as *hyalocytes*, probably they perform the function of macrophages. It shows a surface condensation known as *hyaloid membrane*. The anterior hyaloid passes on the posterior aspect of the lens and the posterior on the internal limiting membrane of the retina.

Vitreous shows a number of ageing changes. Vitreous degeneration, liquefaction, vitreous detachment and shrinkage may occur with advancement of age. The homogeneous vitreous becomes coarse with the age.

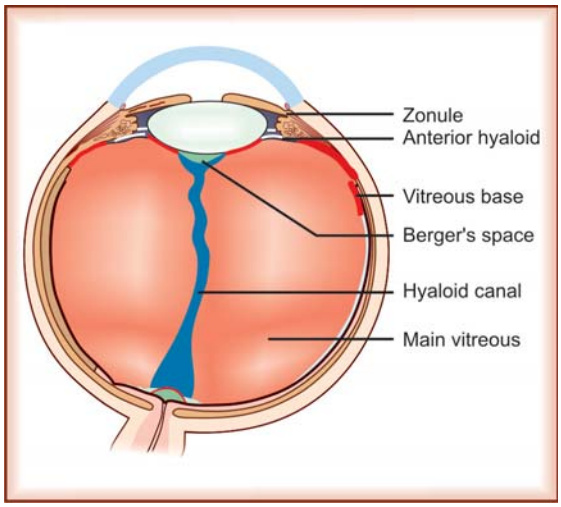

Fig. 17.1: Diagram showing various parts of vitreous and its attachments indicated in red

DISEASES OF THE VITREOUS

Vitreous Opacities

Although vitreous is a transparent hydrophilic gel, a few black specks floating before the eye may be seen by normal persons. They are called *muscae volitantes* and of no clinical significance. They are not visible objectively with an ophthalmoscope. The patient should be reassured and advised to ignore them.

Multiple or large vitreous opacities interfere with the vision and are visible with an ophthalmoscope and on slit-lamp. They are often found

in degenerative and inflammatory conditions of the eye such as myopia, pars planitis and chorioretinitis. In myopia the vitreous loses its gel form and becomes fluid and some of the coagulated gel takes the form of threads and flakes. In pars planitis and retinochoroiditis, leukocytes and fibrinous exudates are released in the vitreous causing its turgescence. Large vitreous floaters are also found after hemorrhage in the vitreous.

Asteroid Hyalosis

Multiple, white round bodies (Fig. 17.2) may be found scattered in the vitreous gel of elderly persons. They are composed of calcium-containing phospholipids and represent asteroid hyalosis (asteroid bodies). They produce little symptoms and occur often unilaterally. Asteroid bodies are adherent to the vitreous structure. Diabetes mellitus and hypertension may be related with them. The opacities may vary in size and are usually unaffected by gravity. They seldom affect the vision, but may cause difficulty in fundus examination. Asteroid bodies causing impairment of vision may be dealt with bimanual vitrectomy.

Synchysis Scintillans

Numerous, yellowish-white, crystalline shining bodies may be found floating in the fluid vitreous in synchysis scintillans. They are composed of cholesterol crystals and occur in eyes that had suffered from vitreous hemorrhage or uveitis. The patient often narrates the appearance of a shower of golden crystals before his eyes on ocular movements. Ordinarily the crystals in synchysis scintillans sink to the bottom of the vitreous cavity. They can be differentiated from asteroid hyalosis on the points listed in Table 17.1.

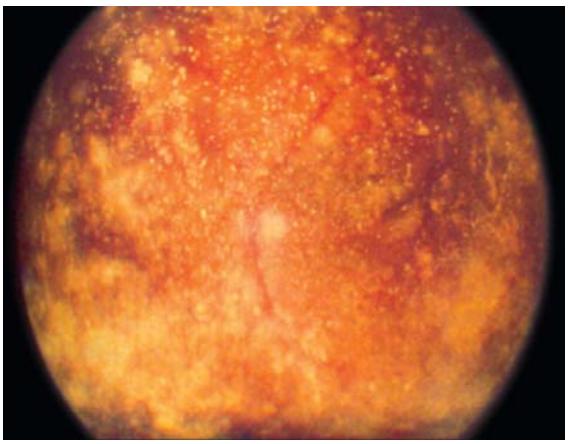

Fig. 17.2: Asteroid hyalosis

Table 17.1: Differentiation between asteroid hyalosis and synchysis scintillans

Features	*Asteroid hyalosis*	*Synchysis scintillans*
Laterality	Unilateral	Bilateral
Composition	Calcium-containing phospholipids	Cholesterol crystals
Symptoms	Mostly symptom-free	Golden crystals before the eye
State of vitreous	Gel	Fluid
Attachments to vitreous	Adherent	Free
Gravity	Unaffected by gravity	Affected by gravity, settles at bottom

Amyloidosis

The heredofamilial amyloidosis is associated with vitreous opacities. The disease is transmitted as an autosomal dominant trait. The ocular features include proptosis, ophthalmoplegia, retinal hemorrhages, cotton-wool spots, exudates and perivasculitis. Both eyes are often involved. The vitreous opacities are classically linear with footplate attachments to the retina and posterior lens surface. They generally cause severe visual impairment. The intravitreal amyloid deposits can be removed by vitrectomy with guarded prognosis.

Sometimes, developmental opacities may be found in the vitreous. They are often located in Cloquet's canal (primary vitreous) and represent remnants of the distal end of hyaloid artery.

Degeneration and Detachment of Vitreous

Vitreous gets degenerated when its gel structure is disrupted. As one gets older the hyaluronic acid concentration decreases, depriving the collagen fibers of their support. Besides senile vitreous degeneration, ocular trauma, high myopia, proliferative diabetic retinopathy and chorioretinitis may also cause vitreous degeneration and fluidity. The condition is diagnosed by the presence of free-floating opacities in the vitreous on slit-lamp examination or ophthalmoscopy. The eye with fluid vitreous runs a risk of complications if intraocular surgery is undertaken.

The liquefied vitreous gains access to the retrohyaloid space, through a hole in the thinner posterior vitreous cortex, and separates the posterior vitreous from the internal limiting membrane of retina. This causes collagen meshwork to collapse and move forward, a phenomenon known as *posterior vitreous detachment* (PVD). It can be asymptomatic or symptomatic. Floaters, flashes of light (photopsia), usually towards the temporal visual field on ocular movements, and diminution of vision due to vitreous hemorrhage are common symptoms of complicated PVD.

A ring of glial tissue, *Weiss ring*, is torn from the optic nerve head which is indicative of PVD over the disk. The detached vitreous may cause dynamic traction on the retina during ocular saccades resulting in retinal tear formation and subsequent detachment. Therefore, all the cases with a history of floaters or photopsia should be thoroughly examined for retinal tears or vitreous hemorrhage. Prophylactic barrage laser photocoagulation or cryopexy of retina is indicated if a break is detected.

Vitreous Bands and Membranes

Vitreous bands (Fig. 17.3) and membranes may develop after the detachment of the posterior vitreous or following massive hemorrhage in the vitreous. They originate from the endothelial cells of the capillaries or from hyalocytes. A sheet of thin tissue may cover the inner retinal surface, the *epiretinal membrane*. Contraction of the membrane or the band can produce a macular pucker or detachment of the retina.

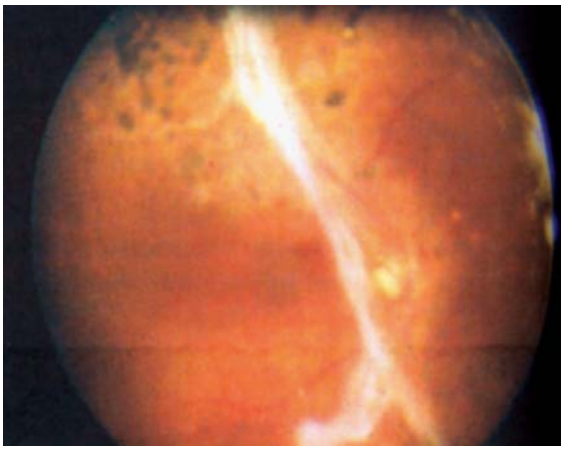

Fig. 17.3: Bands in vitreous

Persistent Hyperplastic Primary Vitreous

Persistent hyperplastic primary vitreous (PHPV) is a rare developmental disorder of the vitreous due to persistence of fetal vasculature. The condition is usually unilateral and frequently seen in a microphthalmic eye. Typically, a white reflex in the pupil is noticed. The presence of a retrolental mass with long extended ciliary processes is characteristic of the *anterior type of PHPV*. Later, it contracts and pulls the ciliary processes inwards. It may be associated with cataract, glaucoma and vitreous hemorrhage.

The *posterior type of PHPV* is less common. It is characterized by a persistent hyaloid system associated with a prominent retinal fold or a stalk extending to the peripheral retina from the optic disk. It may be associated with retinal detachment, pigmentary changes in the choroid and a pale optic disk.

Hemorrhage in the Vitreous

Etiology

Hemorrhage into the vitreous cavity may occur due to various causes, the important ones are as follows:

1. Trauma—blunt or penetrating
2. Proliferative retinopathy:
   a. Diabetic retinopathy
   b. Retinal vein occlusion
   c. Eales' disease
   d. Sickle-cell retinopathy
3. Blood dyscrasias—hemophilia, purpura
4. Posterior vitreous detachment with collapse
5. Retinal breaks with or without detachment.

Clinical Features

The vitreous hemorrhage may be found either in the subhyaloid space or in the vitreous cavity or, sometimes, in both. The subhyaloid blood moves with gravity and appears boat-shaped because it remains unclotted for a long time. When blood in the vitreous cavity clots, it becomes a white opaque mass.

Sudden onset of floaters, diminution of vision or near complete loss of vision are the common symptoms. All cases of vitreous hemorrhage should be carefully examined using an indirect ophthalmoscope. Ultrasonography is particularly helpful in confirming the diagnosis.

Complications

Recurrent vitreous hemorrhages may lead to degeneration of vitreous, tractional retinal detachment, hemolytic or ghost cell glaucoma and hemosiderosis bulbi.

Treatment

Some of the cases of vitreous hemorrhage show significant improvement by bed rest, eye patching and elevation of the head-end of the bed. These methods help prevent dispersion of blood into the vitreous gel. A two monthly follow-up is desirable to assess the progress in clearance of the vitreous hemorrhage. If the blood does not absorb within six months and the patient has visual acuity less than 6/60 or if vitreous hemorrhage is associated with retinal detachment, vitrectomy is indicated.

Parasites in the Vitreous

Parasites in the vitreous are rare. Cysticercus in the vitreous may be seen ophthalmoscopically as a pearly, translucent mass (Fig. 17.4) showing peristaltic movements. It may occur alone or may be associated with subretinal cysticercus cyst. The cyst has to be removed by pars plana vitrectomy (See video).

Prepapillary Vascular Loops

Prepapillary vascular loops are normal retinal vessels, mainly arterial, that extend in *Bergmeister's papilla* before returning to the optic disk.

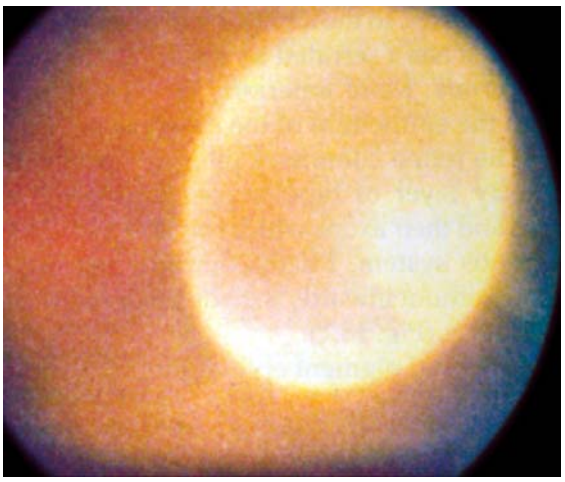

Fig. 17.4: Intravitreal cysticercosis

Bergmeister's papilla is the posterior remnant of tunica vasculosa lentis that appears as a stalk of fibroglial tissue emanating from the optic nerve head into the vitreous. The prepapillary vascular loops may cause vitreous hemorrhage and branch retinal artery occlusion.

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CHAPTER

18 Diseases of the Retina

ANATOMY

The innermost and highly developed layer of the eyeball is known as *retina*. In fact, the retina is a part of the brain and develops from the optic vesicle, an outgrowth from the forebrain. The outer wall of the vesicle forms the retinal pigment epithelium and the inner, the neurosensory retina. The retina, a thin transparent membrane, lies between the choroid and the hyaloid membrane of vitreous. It extends from the optic disk to the anterior end of the choroid where it has a serrated termination known as *ora serrata*. More anteriorly it is continuous with the epithelium of the ciliary body.

The retina comprises photoreceptor cells, a relay layer of bipolar cells and ganglion cells and their axons that run into the central nervous system. Microscopically, the retina from without inwards is made up of following ten layers (Fig. 18.1).

1. Retinal pigment epithelium
2. Layers of rods and cones
3. External limiting membrane
4. Outer nuclear layer
5. Outer plexiform layer
6. Inner nuclear layer
7. Inner plexiform layer
8. Ganglion cell layer
9. Nerve fiber layer, and
10. Internal limiting membrane.

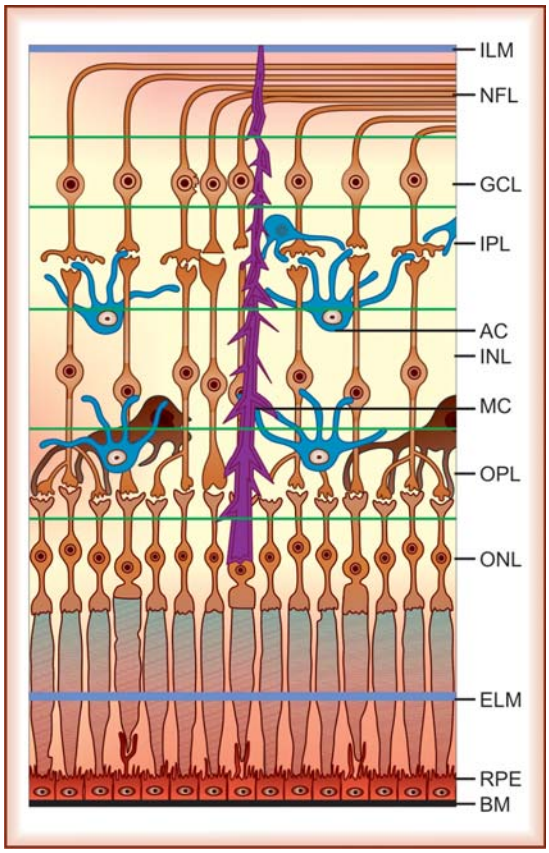


Fig. 18.1: Diagram showing various layers of retina. ILM, Internal limiting membrane; NFL, Nerve fiber layer; GCL, Ganglion cell layer; IPL, Inner plexiform layer; AC, Amacrine cell; INL, Inner nuclear layer; MC, Muller's cell; OPL, outer plexiform layer; ONL, Outer nuclear layer; LRC, Layer of rods and cones; ELM, External limiting layer; RPE, Retinal pigment epithelium; BM, Bruch's membrane.

The rods and cones are the end organs of vision and are photosensitive (Fig. 18.2). The various layers of the retina are bound together by neuroglia. There are well-developed vertical fibers of Muller which have supportive as well as nutritive functions. The internal limiting membrane separates the retinal nerve fiber layer from the vitreous, while the external limiting membrane is perforated by the rods and cones.

At the posterior pole there is a circular area which appears darker than the surrounding retina—*macula lutea*. It has a diameter of 5.5 mm, the horizontal diameter is slightly greater than the vertical. The center of the macula is marked by a depression called *fovea centralis*. It is approximately 2 disk diameters away from the temporal margin of the optic disk and about 1 mm below the horizontal meridian. The fovea centralis is a highly differentiated spot where only cones are present and the other layers of the retina are almost absent (Fig. 18.3). It is the most sensitive part of the retina and has the maximal visual acuity.

The blood supply of inner layers of retina comes from the central retinal artery and its branches. These arteries are end arteries and, as such, they do not anastomose excepting in the neighborhood of the lamina cribrosa. The outer layers of retina up to the outer nuclear layer get their nourishment by diffusion from the choriocapillaris. The outer plexiform layer is nourished by diffusion from the choriocapillaris as well as by the retinal vascular system. The venous drainage of inner layers of the retina is through the retinal veins that do not exactly follow the course of the arteries. These veins and the central retinal vein, which follows the course of corresponding artery, ultimately join the cavernous sinus. The outer retinal layers are drained by the vortex veins.

DISEASES OF THE RETINA

Congenital and developmental, vascular, inflammatory, degenerative, infiltrative and neoplastic

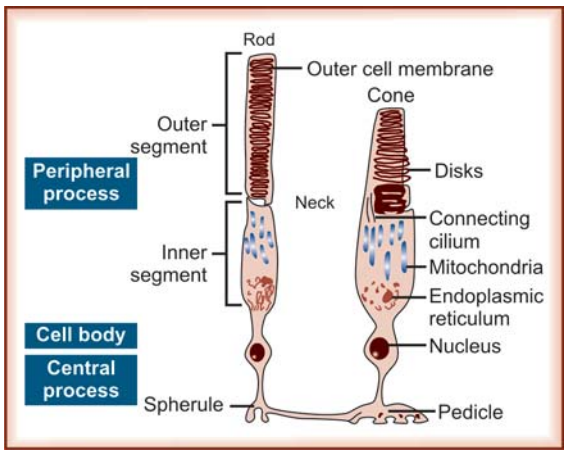


Fig. 18.2: Diagram showing a rod and a cone

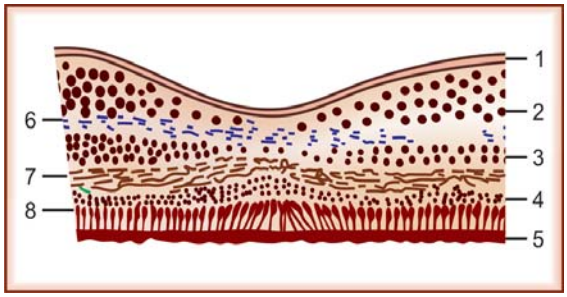


Fig. 18.3: Transverse section of macula
1. Internal limiting membrane and nerve fiber layer, 2. Ganglion cell layer, 3. Inner nuclear layer, 4. Outer nuclear layer, 5. Pigment epithelium, 6. Inner plexiform layer, 7. Outer plexiform layer, 8. Layer of cones

affections of the retina are not uncommon. Since neurosensory retina is a delicate and loosely attached membrane, it is prone for separation or detachment.

CONGENITAL AND DEVELOPMENTAL ANOMALIES OF THE RETINA

Coloboma of the Retina

A typical coloboma of the retina associated with coloboma of the choroid is situated downwards and inwards. The retina fails to develop in the region due to non-closure of the optic fissure. The

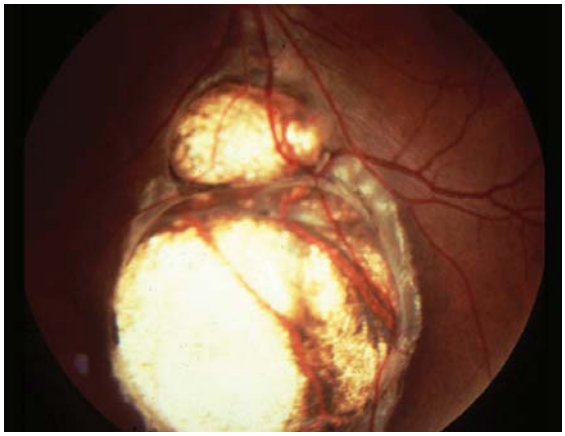


Fig. 18.4: Typical coloboma of retina and choroid

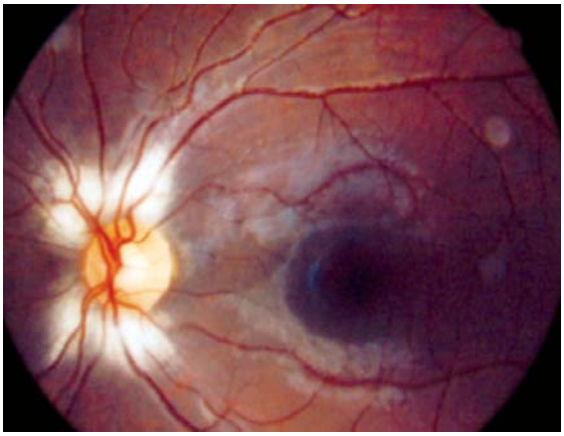


Fig.18.5: Medullated nerve fibers

coloboma appears as an oval depressed defect with rounded apex towards the disk (Fig. 18.4). A few vessels may be seen over the surface and the edges contain some pigments.

Opaque Nerve Fibers

The myelination of the optic nerve progresses from the brain towards the periphery and stops at the lamina cribrosa. It is usually completed shortly after birth. However, occasionally some of the nerve fibers near the optic disk become myelinated that on ophthalmoscopic examination appear as white patches with feathery margins covering the retinal vessels and are termed as *opaque nerve fibers* or *medullated nerve fibers* (Fig. 18.5). They should be clinically differentiated from cotton-wool spots.

Congenital Pigmentation of the Retina

It is not rare to see small, oval, gray or black polygonal spots in the retina lying below the vessels which are labeled as *congenital pigmentation of the retina*. They occur due to abnormal heaping of the retinal pigment epithelium.

VASCULAR DISORDERS OF THE RETINA

The retina is richly supplied by blood vessels and, therefore, it is not uncommon to observe its involvement in inflammatory and systemic diseases.

Hyperemia of the Retina

Hyperemia of the retina may occur due to either inflammatory lesions of retina and choroid or venous obstruction. The venous hyperemia is characterized by dilatation and tortuosity of the veins and seen in central retinal vein occlusion, papilledema, congestive cardiac failure and congenital heart diseases. Retinal hyperemia is an important feature of polycythemia vera and leukemia.

Anemia of the Retina

Anemia of the retina is commonly found in central retinal artery occlusion, quinine poisoning and spasm of the retinal arteries due to toxemia of pregnancy. It can be a local expression of profuse

hemorrhage, severe anemia and arteriosclerosis. Generalized pallor of the fundus as well as the optic disk, attenuation of retinal vessels and white-centered hemorrhages (*Roth's spots*) are the characteristic features. Usually the retinal vessels show constriction in hyperoxemia (oxygen concentration in the blood is high) and dilatation in hypoxemia.

Edema of the Retina

The retinal edema may be diffuse or localized. The diffuse retinal edema renders the bright red retina a dull pale sheen with white streaks along the course of blood vessels.

A localized edema in the macular region presents a characteristic star owing to the formation of radiating folds following accumulation of transudate in Henle's layer. A *macular star* may be found in hypertensive retinopathy and papilledema. A milky-white cloudiness develops at the posterior pole due to edema as a result of blow on the globe, such an edema is known as *commotio retinae* or *Berlin's edema* (Fig. 18.6). The central vision is usually diminished and later pigmentary changes appear at the macula causing severe visual loss. Sometimes, the macular edema disappears and vision is restored.

There occurs an accumulation of fluid in Henle's layer and inner nuclear layer of the macula in cases of *cystoid macular edema* (CME). Fluorescein angiography demonstrates abnormal permeability of perifoveal capillary network. Cystoid macular edema may develop following ocular surgeries like cataract extraction (Irvine-Gass syndrome), filtration surgery, retinal reattachment surgery, vitrectomy and photocoagulation. It is less frequent following extracapsular lens extraction than the intracapsular cataract extraction. Other causes of CME include diabetic retinopathy, retinal vein occlusion, uveitis, retinitis pigmentosa and topical indiscriminate use of prostaglandin analog and epinephrine in the postoperative phase.

Cystoid macular edema shows a loss of foveal reflex and edema of the macula with multiple cystoid spaces giving a honeycomb appearance on slit-lamp biomicroscopy using 90 D lens. Fluorescein angiography (FA) is diagnostic which shows an area of hyperfluorescence giving a *flower-petal appearance* in the late phase of angiogram (Fig. 18.7). Generally, visual prognosis of CME is good and in most of the cases it resolves spontaneously.

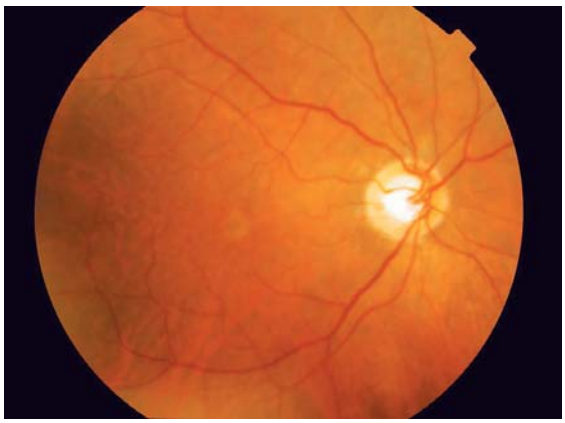


Fig. 18.6: Commotio retinae (*Courtesy:* Dr Sanjay Thakur, Nataraj Eye Centre, Varanasi)

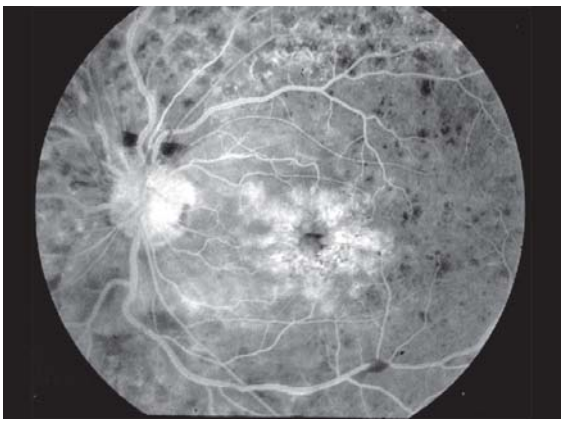


Fig. 18.7: FA of cystoid macular edema showing flower-petal appearance

Development of cystoid macular edema must be prevented. Systemic and topical non-steroidal anti-inflammatory drugs (NSAIDs) should be used preoperatively in all the cases of intraocular surgery. Postoperatively topical NSAIDs are continued for 3 to 6 months in susceptible patients. Associated diseases must be treated to reduce the incidence of CME. If CME develops, topical, periocular and systemic steroids are given for early resolution and prevention of irreversible retinal damage. Chronic form of CME may be treated by grid photocoagulation with variable visual results.

Retinal Hemorrhages

The retinal hemorrhages are either intraretinal (within the tissue) or preretinal. The hemorrhages assume a characteristic appearance according to their location, conforming to the anatomical peculiarities of the layer in which they lie. The *superficial hemorrhages* lie in the nerve fiber layer and assume striate or flame-shaped appearance. *Intraretinal hemorrhages* are round or irregular as they lie in the deeper layers. *Preretinal* or *subhyaloid hemorrhages* commonly occur near the macula (Fig. 18.8). Initially they are large and round, but soon become hemispherical due to the sedimentation of erythrocytes. Large hemorrhages tend to involve the vitreous rendering it opaque, so that the red fundus glow is lost. Recurrent hemorrhages are absorbed very slowly and occasionally induce the proliferation of fibrous tissue—*retinitis proliferans*. The contraction of fibrous tissue may cause tractional retinal detachment. Retinal hemorrhages can occur in a wide variety of conditions such as trauma, hypertensive and diabetic retinopathies, occlusion of central or branch retinal vein and blood dyscrasias.

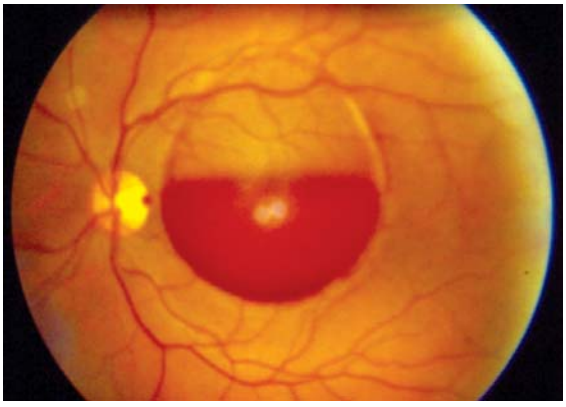


Fig. 18.8: Subhyaloid hemorrhage

Coats' Disease

Coats' disease or exudative retinopathy of Coats is an uncommon uniocular condition mostly found in young males. It is characterized by telangiectatic blood vessels, multiple small aneurysms and varying amount of yellowish-white exudates and hemorrhages near or temporal to the fovea. The lesion is usually raised and may cause exudative detachment of retina, cataract and glaucoma. Early lesions may be treated by laser photocoagulation.

Retinopathy of Prematurity

Retinopathy of prematurity (ROP) is a proliferative retinopathy of premature (gestational age of 28 weeks or less) low weight (less than 1500 gm) infants. ROP is caused by excessive oxygenation of premature babies during first few weeks of life. The oxygenation causes obliteration of premature retinal blood vessels followed by fibrovascular proliferation. Besides exposure to excessive concentration of oxygen, low birth weight and prematurity increase the risk of developing the disease.

Retinopathy of prematurity is a bilateral asymmetrical disease. According to the location of the disease, ROP is usually classified and documented in 3 zones (Fig. 18.9): *zone I* encompasses the posterior retina within a 60° circle centered on the optic nerve, *zone II* involves the zone I and the nasal ora serrata anteriorly, and *zone III* includes the remaining temporal peripheral retina outside zones I and II.

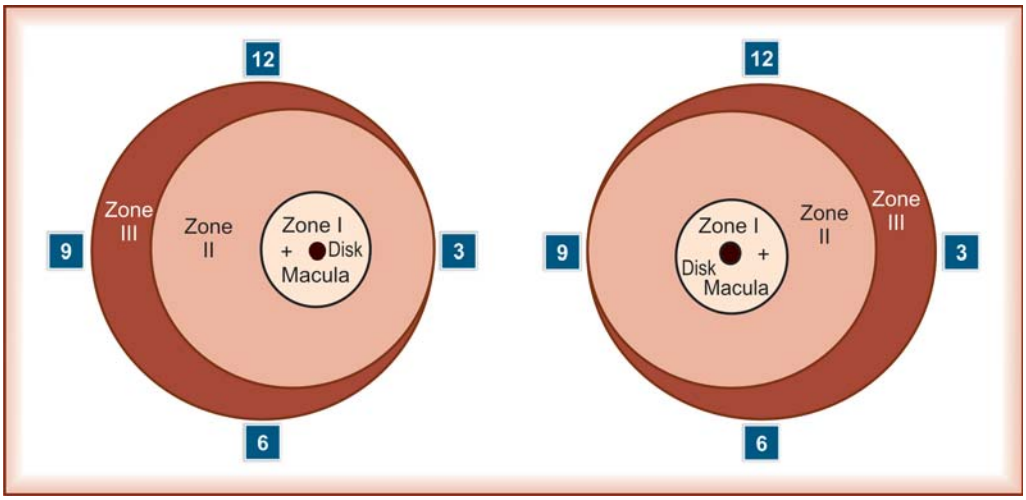


Fig. 18.9: ROP zones
(*Courtesy*: Prof. RV Azad, Dr RP Centre, New Delhi)

The course of ROP may be divided into 5 stages.

Stage 1: A thin irregular grayish-white demarcation line is seen separating the avascular peripheral retina from the vascular posterior retina.

Stage 2: The demarcation line develops into a ridge with vascular tufts.

Stage 3: The ridge is associated with extraretinal fibrovascular proliferation and hemorrhages in the retina and the vitreous.

Stage 4: A subtotal tractional retinal detachment occurs. In stage 4A the detachment is extrafoveal and in stage 4B the fovea is involved.

Stage 5: It is characterized by total retinal detachment giving an amaurotic cat's eye reflex (Fig. 18.10).

Retinopathy of prematurity is differentiated from retinoblastoma by a positive history of prematurity, low weight at birth and oxygen therapy.

At least 2 detailed dilated fundus examinations using an indirect ophthalmoscope are recommended for all infants with a birth weight of less than 1500 grams or with a gestational age

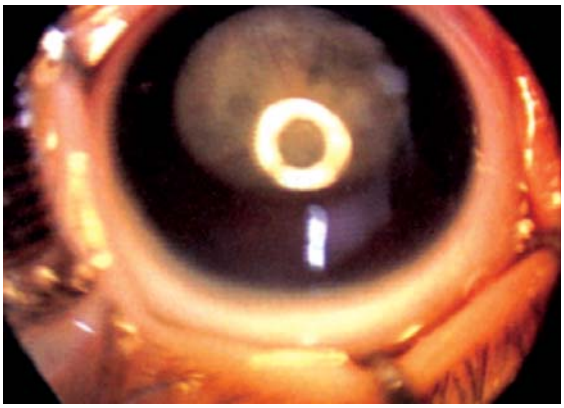


Fig. 18.10: Retinopathy of prematurity Stage 5
(*Courtesy*: Prof. RV Azad, Dr RP Centre, New Delhi)

of 28 weeks or less in order to screen for retinopathy of prematurity. The first examination is performed between 4 and 6 weeks postnatally. The child should be followed up every 1 to 2 weeks until the retina gets fully vascularized.

ROP shows spontaneous regression in 85% of eyes. The best preventive measure for ROP is to avoid low birth weight and controlled supplemental oxygen therapy using a pulse oximeter.

Laser photocoagulation of the avascular retina is preferred over cryoablation up to stage 3 of the disease. Scleral buckling with or without vitrectomy is indicated in eyes with stage 4 ROP. Stage 5 ROP is managed by advanced bimanual vitrectomy with sectioning of the bands and membrane peeling.

Central Retinal Artery Occlusion

Etiology

The central retinal artery is occluded almost always at the lamina cribrosa due to its narrowing. The obstruction in young people may be due to the spasm of vessels seen in toxemia of pregnancy and quinine toxicity, while in advanced age group it is associated with arteriosclerosis and hypertension. An embolic occlusion is rare, the embolus may come from endocarditis. The obstruction of central retinal artery may be found in giant cell arteritis, systemic lupus erythematosus, polyarteritis nodosa and Takayasu's disease. Occasionally, the occlusion may occur due to increased intraocular pressure as seen in acute angle-closure glaucoma or excessive pressure on the globe during retinal reattachment surgery or neurosurgical procedures.

Clinical Features

The eye may become suddenly blind without any pain. The ophthalmoscopic examination reveals a characteristic picture. The retinal arteries appear thread-like, while there is no change in the caliber of retinal veins. The retina becomes ischemic, edematous and milky-white (Fig. 18.11). There occurs a striking *cherry-red spot* at the macula as the choriocapillaris shine against the ischemic white background of macular edema. If the occlusion is incomplete, a slight pressure on the globe may present a *cattle-truck phenomenon*, the segmented blood column moves in a jerky way in the veins sometimes in the normal direction of blood flow and sometimes in the opposite.

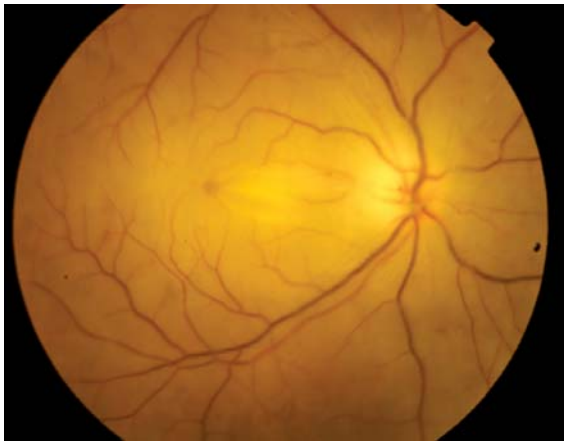


Fig. 18.11: Central retinal artery occlusion (*Courtesy*: Prof. YR Sharma, Dr RP Center for Ophthalmic Sciences, New Delhi)

In some cases, despite a complete central retinal artery occlusion (CRAO) some degree of central vision is retained due to the presence of a cilioretinal artery that supplies the macular region. Rarely, only the cilioretinal artery becomes blocked.

When the central retinal artery blockage remains for more than 90 minutes, the retina undergoes atrophic changes with serious visual loss. Later the edema clears up, the retina regains its transparency and near normal sheen mostly owing to the viability of its outer layers which get their nourishment from the choroid. The eye loses useful vision due to optic atrophy, and the direct pupillary reaction to light becomes absent.

Rubeosis iridis may develop in a small percentage of eyes with obstruction of the central retinal artery that may be complicated by neovascular glaucoma. It occurs sooner after arterial occlusion (mean period of 4-5 weeks) than venous occlusion.

Differential Diagnosis

The patients of CRAO must be differentiated from other causes of cherry-red spot at the macula that include commotio retinae, Tay-Sachs disease, Sandhoff disease, generalized gangliosidosis,

Niemann-Pick disease, sialidosis and galacto-sialidoses.

Treatment

Treatment of central retinal artery occlusion is very unsatisfactory. If the patient reports early, attempts should be made to restore the retinal circulation by massaging the globe intermittently for at least 15 minutes. Attempts should be made to lower the intraocular pressure by giving intravenous acetazolamide (500 mg) or retrobulbar anesthesia or by doing anterior chamber paracentesis. Inhalation of amyl nitrate or a mixture of 5 percent carbon dioxide and 95 percent oxygen have been tried with varying results. If giant cell arteritis is the underlying cause, high dosage of systemic steroids is instituted. Neovascularization of iris following central retinal artery occlusion is dealt with panretinal photocoagulation.

Branch Retinal Artery Occlusion

When a branch of the retinal artery is obstructed (Fig. 18.12) the sector supplied by it is affected associated with a permanent sectorial visual field defect. The most common site of obstruction of a branch retinal artery is superotemporal.

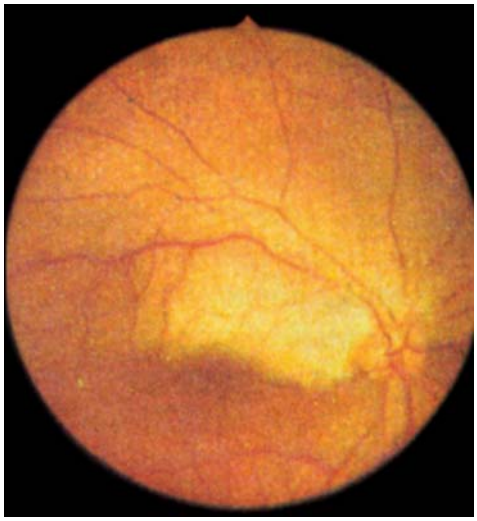


Fig. 18.12: Branch retinal artery occlusion (*Courtesy*: Mr S Kanagami, Tokyo)

Etiology

The most common cause of obstruction of a branch retinal artery is thromboembolic phenomenon. The emboli can be from carotid arteries containing cholesterol (*Hollenhorst plaques*), arteriosclerotic vessels consisting of platelet-fibrin, or cardiac valvular diseases comprising calcium. Other causes include fat emboli from fractures of long bones, septic emboli form infective endocarditis and talc emboli in drug abusers. Sickle-cell anemia, coagulation disorders, trauma, migraine, and oral contraceptives may also cause branch retinal arterial occlusion.

Treatment

The management of branch retinal artery occlusion includes treating the underlying systemic condition and digital ocular massage with the aim of dislodging the embolus to a peripheral vessel in the retina.

Central Retinal Vein Occlusion

Etiology

Central retinal vein occlusion (CRVO) is a unilateral condition usually occurring in elderly people with cardiovascular disorders. The obstruction often occurs at or just behind the lamina cribrosa due to thrombus formation where the vein shares a common sheath with the central retinal artery. The central retinal vein may get occluded in periphlebitis retinae, sarcoidosis, Behçet's disease and orbital cellulitis. Approximately, one-third cases of venous obstruction have associated open-angle glaucoma. Diabetes, hypertension, leukemia, sickle-cell anemia, polycythemia vera and multiple myeloma are considered as risk factors for the obstruction. Oral contraceptives and diuretics are also blamed for causing venous occlusion.

Types

The central retinal vein occlusion occurs in two forms.

1. *Ischemic central retinal vein occlusion* presenting as hemorrhagic retinopathy and accompanied by retinal ischemia, and
2. *Nonischemic central retinal vein occlusion* presenting as venous stasis retinopathy without ischemia.

Ischemic Central Retinal Vein Occlusion

Clinical Features

The ischemic CRVO occurs in the region of lamina cribrosa. It causes painless loss of vision not so sudden as found in the central retinal artery occlusion. Some patients complain of blackouts and red vision (erythropsia). There is a relative afferent pupillary defect in the affected eye. Fundus examination reveals engorged, tortuous retinal veins, edema of the disk and retina, flame-shaped and blot hemorrhages in all four quadrants of the retina, and cotton-wool spots. The hemorrhages are so extensive that the entire retina appears covered with them (Fig. 18.13). The retinal arterioles show sclerotic changes. Fluorescein angiography reveals areas of capillary non-perfusion. The visual prognosis in ischemic central retinal vein occlusion is usually poor due to the development of neovascular glaucoma and macular complications.

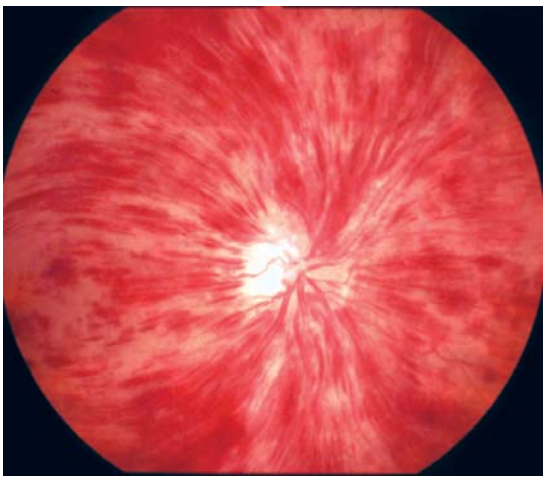


Fig. 18.13: Central retinal vein occlusion (*Courtesy*: Mr S Kanagami, Tokyo)

Recurrent vitreous hemorrhages are frequent in classical CRVO due to neovascularization of the retina and the optic disk. Retina undergoes pigmentary and atrophic changes. Cystoid degeneration of macula, optic atrophy and hemorrhagic or neovascular glaucoma are serious complications of CRVO. The hemorrhagic glaucoma is also known as *90-day glaucoma* because it ensues nearly 3 months after the episode of occlusion.

Nonischemic Central Retinal Vein Occlusion *(*Lyle-Wybar Syndrome*)*

Mild central retinal vein occlusion occurs in young persons unaffected by systemic vascular disorder.

Clinical Features

Nonischemic central retinal vein occlusion is characterized by dilated tortuous veins, a few intraretinal hemorrhages and cotton-wool spots, and features of retinal perfusion on fluorescein angiography. Visual acuity in nonischemic occlusion is better than that in ischemic CRVO.

Treatment

Special attention is given to manage the associated systemic conditions in cases of CRVO. There is no satisfactory treatment for an ischemic central retinal vein occlusion. Panretinal photocoagulation or, if media are hazy, anterior retinal cryopexy may prevent neovascular glaucoma in patients with iris neovascularization.

Oral corticosteroids may be helpful in some cases of nonischemic CRVO. Retinal vein cannulation with tissue plasminogen activator (tPA) infusion and decompression of central retinal vein by radial optic neurotomy (incising the posterior

scleral ring) have been tried with variable results. Injection of triamcinolone acetonide intravitreally in patients of CRVO has been claimed to reduce the macular edema.

Cystoid macular edema may be dealt with grid laser photocoagulation. However, Central Vein Occlusion Study had shown that grid laser treatment only hastens the resolution of macular edema in treated eyes without improving the visual acuity as compared to control eyes.

Branch Retinal Vein Occlusion

Branch retinal vein occlusion (BRVO) is common in the superotemporal branch due to numerous crossings by the artery. It is commonly associated with systemic hypertension, cardiovascular disease, obesity and raised intraocular pressure. The visual impairment may not be noticed in branch vein occlusion until macula is involved which may be in the form of edema, hemorrhage or perifoveal capillary occlusion. In acute branch vein occlusion, flame-shaped hemorrhages, edema and cotton-wool spots are confined to the area drained by the vein (Fig. 18.14).

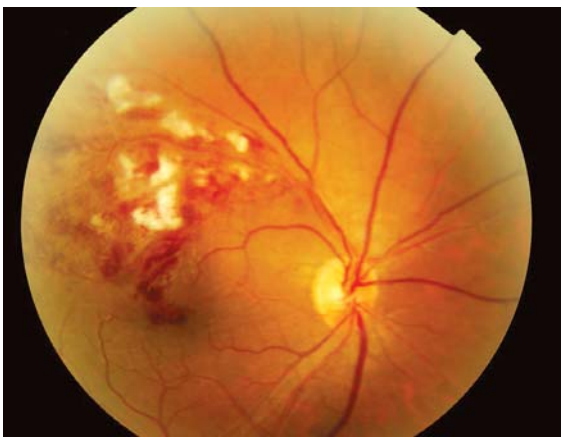


Fig. 18.14: Branch retinal vein occlusion
(*Courtesy:* Dr Sanjay Thakur, Nataraj Eye Centre, Varanasi)

Treatment

The underlying predisposing conditions must be attended properly. Photocoagulation is advocated for macular edema and retinal neovascularization. At least 3-month time is given for the spontaneous resolution of edema and hemorrhages before instituting the laser therapy. Pars plana vitrectomy with or without arteriovenous sheathotomy is done for recurrent non-clearing vitreous hemorrhage.

VASCULAR RETINOPATHIES

Retinal manifestation of a systemic vascular disorder is termed as *retinopathy*. It is usually bilateral and noninflammatory in origin. Diabetes, arteriosclerosis, hypertension, nephritis, toxemia of pregnancy, blood dyscrasias and systemic lupus erythematosus produce characteristic vascular retinopathies.

Diabetic Retinopathy

Diabetic retinopathy (DR) is a major cause of blindness in elderly subjects, and develops frequently in long-standing cases of diabetes mellitus especially of more than 10 years duration.

DR is a microangiopathy involving the retinal precapillary arterioles, the capillary bed and the postcapillary venules. The pathogenesis of diabetic retinopathy includes both microvascular occlusion and leakage. Fundus changes in diabetic retinopathy may be intraretinal, preretinal or vitreal. Diabetic retinopathy is conventionally divided into two broad categories.

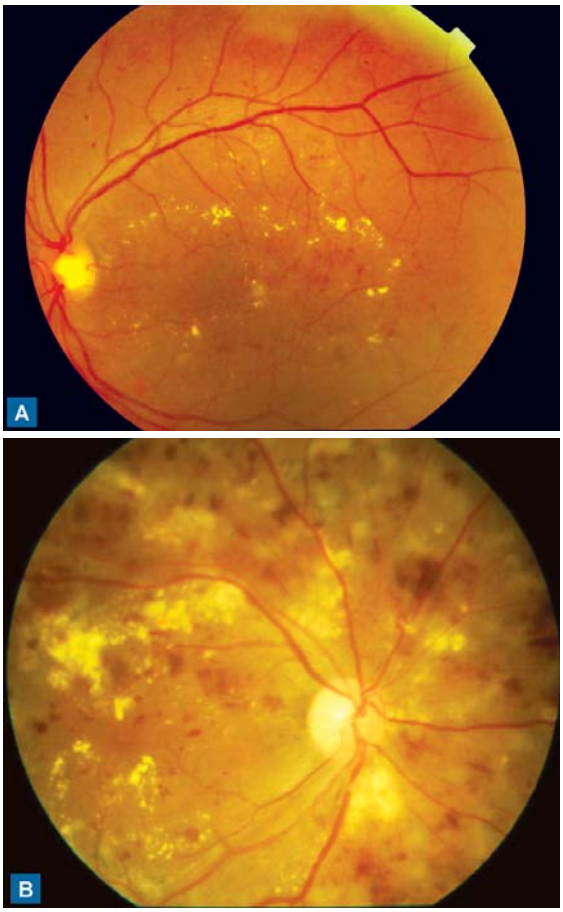
1. Nonproliferative (background) diabetic retinopathy, and
2. Proliferative diabetic retinopathy.

Nonproliferative Diabetic Retinopathy

Nonproliferative diabetic retinopathy (NPDR) is the most common type of diabetic retinopathy

wherein the lesions are intraretinal and confined to the posterior pole. It is characterized by multiple microaneurysms, venous dilatation, hard exudates, dot and blot and flame-shaped hemorrhages and retinal edema. The earliest sign of NPDR is a capillary microaneurysm. The microaneurysms appear as multiple, minute, round, red dots occasionally arranged like clusters of grapes at the ends of paramacular vessels. They are usually associated with yellow-white waxy-looking exudates with crenated margins (Figs 18.15A and B).

The exudates may coalesce to form irregular big plaques at the posterior pole. The retinal veins are engorged, irregularly dilated and tortuous, and may show beading. The venous changes may be the only sign found in juvenile diabetic retinopathy. The arteries may look normal. Cotton-wool spots are seen especially in diabetics with associated hypertension. Deep, round, dot and blot hemorrhages are found scattered in the retina, while flame-shaped hemorrhages originate from large vessels and lie superficially in the nerve fiber layer. The retinal edema is mostly confined to the macular area.



Figs 18.15A and B: Nonproliferative diabetic retinopathy

Severity of NPDR is expressed by *4:2:1 rule* which is characterized by retinal hemorrhages and microaneurysms in 4 quadrants, venous beading in 2 quadrants and intraretinal microvascular abnormalities (IRMA), which represent shunt vessels that run from retinal arterioles to venules bypassing the capillary bed, in 1 quadrant. The presence of any 1 of these features represents severe NPDR while any 2 features indicates very severe NPDR and risk for progression to proliferative diabetic retinopathy.

Diabetic Macular Edema

Diabetic macular edema in NPDR is the most common cause of decreased vision. *Clinically significant macular edema* (CSME) reduces or threatens to reduce the vision. It is best detected by stereoscopic biomicroscopy using 78 or 90 diopter lens. CSME includes: (i) retinal edema at or within 500 microns of the center of the foveal avascular zone (FAZ), (ii) exudates at or within 500 microns of the center of FAZ associated with thickening of the adjacent retina, and/or (iii) retinal edema 1 disk area or larger in size within 1 disk diameter of the center of FAZ.

Diabetic Maculopathy

Diabetic maculopathy may be focal, cystoid or ischemic. In *focal* or *exudative maculopathy*, mild macular thickening and a few hard exudates are seen on slit-lamp biomicroscopy (Fig. 18.16). *Cystoid maculopathy* is characterized by accumulation of

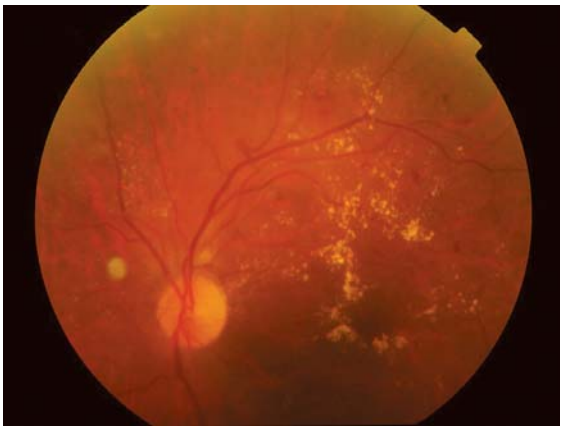


Fig. 18.16: Diabetic maculopathy

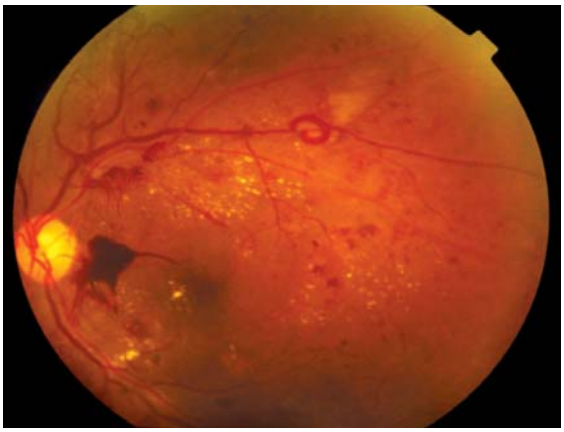


Fig. 18.17: Proliferative diabetic retinopathy

fluid in Henle's layer with microcystic spaces. *Ischemic maculopathy* shows closure of perifoveal capillary net and enlargement of foveal avascular zone on fluorescein angiography. It has the worst visual prognosis.

Proliferative Diabetic Retinopathy

Proliferative diabetic retinopathy (PDR) develops in about 5% of diabetic population. Proliferative changes are a response to the release of vascular endothelial growth factor (VEGF) from ischemic retina. In PDR the changes are preretinal as well as vitreal. Neovascularization of the optic disk (NVD) and neovascularization elsewhere (NVE), posterior detachment and collapse of the vitreous, vitreoretinal fibrovascular bands and vitreous hemorrhage characterize proliferative diabetic retinopathy. Neovascularization is the hallmark of PDR. It occurs on the optic nerve head and along the major temporal vascular arcades (Fig. 18.17). The proliferation of fibrovascular tissue on the surface of the retina and in the vitreous may cause formation of epiretinal membrane and irregular fibrovascular bands, respectively. The contraction of these bands may lead to tractional retinal detachment and blindness.

Treatment

Medical treatment of DR is aimed at prevention of retinopathy. Tight glycemic control is associated with reduction in development of retinopathy. Good metabolic control and proper management of hypertension prevent the progression of DR.

In patients with diabetic maculopathy, fluorescein angiography is performed to detect the treatable lesions. All leaking microaneurysms, 500 microns or more from the center of FAZ, must be treated by focal laser photocoagulation. Recalcitrant cases may be dealt with intravitreal injection of triamcinolone acetonide (4 mg/0.1 ml). Diffuse capillary leak, as seen in cystoid maculopathy, is treated by grid pattern of photocoagulation in which 100 or 200 microns size burns of moderate intensity are placed, one burn width apart, in the macular area sparing the FAZ. Exudative maculopathy is dealt with focal photocoagulation.

Panretinal photocoagulation (Fig. 18.18) is done in cases of PDR with neovascularization. Despite panretinal photocoagulation (PRP) some cases develop repeated hemorrhages, non-clearing vitreous hemorrhage and traction retinal detachment that require pars plana vitrectomy.

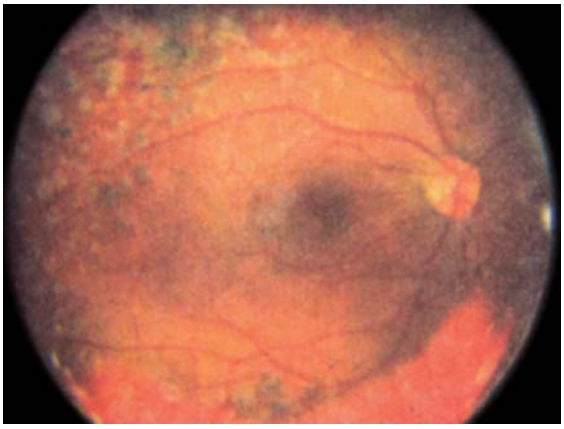


Fig. 18.18: Panretinal photocoagulation in PDR

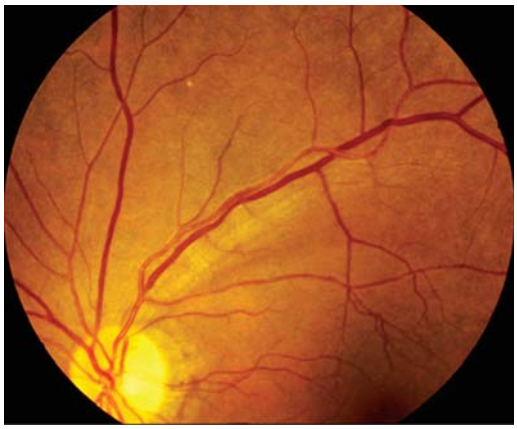


Fig. 18.19: Hypertensive retinopathy grade 2 (*Courtesy*: Mr S Kanagami, Tokyo)

Arteriosclerotic Retinopathy

Arteriosclerotic retinopathy occurs due to senile arteriosclerosis and is commonly associated with hypertension. In the early stage, it is characterized by an increased light reflex, focal attenuation and irregularity in caliber of the retinal arteries. These changes are essentially due to fibrosis and hyalinization of the vessel wall. The changes are indistinguishable from those of hypertension. As the process of sclerosis advances, the vessel wall gradually loses its transparency and the artery assumes a burnished appearance (*copper-wire artery*) or becomes silver-wire. Arteriosclerotic changes are striking at the arteriovenous crossings, and include concealment of the vein under a hardened artery, banking of the vein distal to its arterial crossing (*Bonnet's sign*), tapering of the vein on either side of the crossing (*Gunn's sign*) and right-angled deflection of the vein (*Salus' sign*). Arteriosclerotic retinopathy may be associated with a branch retinal vein occlusion. In addition to classical changes, flame-shaped hemorrhages and hard exudates may be present.

Hypertensive Retinopathy

Hypertension adversely affects the retinal vessels and induces narrowing. The hypertensive narrowing in its pure form can only be seen in young individuals, while in older people arteriosclerotic changes are added upon. Diffuse or focal or mixed arterial narrowing, cotton-wool spots, microaneurysms, flame-shaped hemorrhages, exudates, small branch arteriolar or vein occlusion and optic nerve head edema may be found in moderate to severe hypertensive retinopathy.

Hypertensive retinopathy is classified into five grades according to modified Scheie's classification. It includes the changes of arteriosclerosis also.

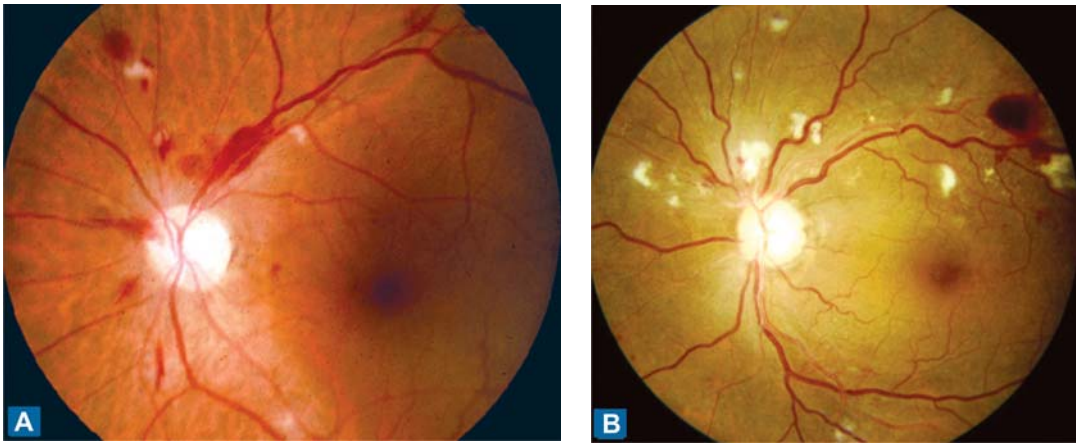
Grade 0: No changes

Grade 1: Visible arteriolar narrowing

Grade 2: Obvious arteriolar narrowing with localized irregularities (Fig. 18.19)

Grade 3: Besides grade 2 changes, there are multiple flame-shaped hemorrhages, cotton-wool spots and/or exudates (Figs 18.20A and B).

Grade 4: It is also known as *malignant hypertension.* In addition to grade 3 changes, the presence of the papilledema (optic disk edema) is an important feature (Fig. 18.21). Papilledema is often accompanied with retinal edema and, in long-standing cases, with macular star (Fig. 18.22).



Figs 18.20A and B: Hypertensive retinopathy grade 3 (*Courtesy*: Mr S Kanagami, Tokyo)

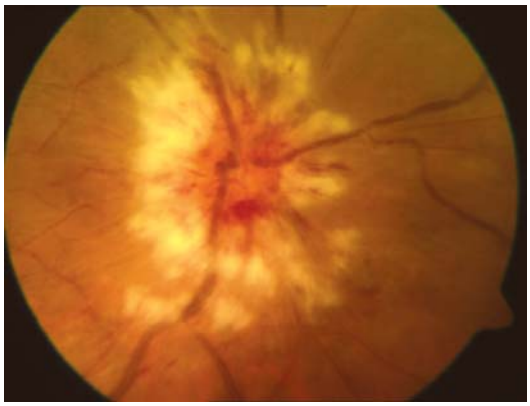


Fig. 18.21: Hypertensive retinopathy grade 4 (*Courtesy*: Dr Tarun Sharma, Sankara Nethralaya,Chennai)

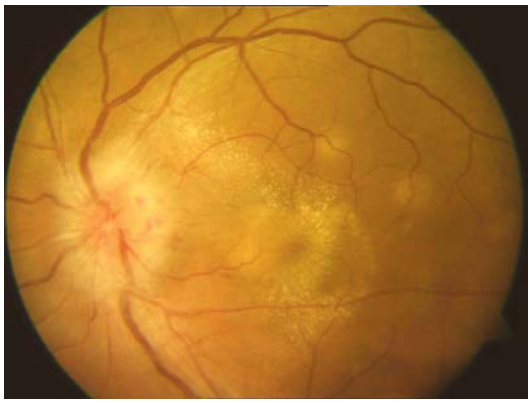


Fig. 18.22: Hypertensive retinopathy grade 4 with macular star (*Courtesy*: Dr Tarun Sharma, Sankara Nethralaya, Chennai)

Hypertensive retinopathy may cause loss of vision due to macular hemorrhage, retinal edema and hard exudates. These changes are related to poor macular capillary perfusion. The grading of retinopathy has prognostic significance. It also determines the efficacy of treatment. Even severe cases of hypertensive retinopathy (grade 3) are reversible, but malignant hypertensive retinopathy may take several months to resolve. The visual recovery may not be complete in these patients even if systemic arterial hypertension is adequately controlled.

Renal Retinopathy

Renal retinopathy develops in cases of chronic diffuse glomerulonephritis associated with systemic hypertension and rarely in acute nephritis. It often causes diminution of vision. The ophthalmoscopic picture is more or less identical to that of malignant hypertension. The retinal edema is marked and the optic nerve head may be swollen. Retinal arteries are grossly attenuated. Numerous flame-shaped hemorrhages are scattered over the fundus (Fig. 18.23). Cotton-wool spots and hard exudates are frequent and macula presents a star figure. Longstanding cases of

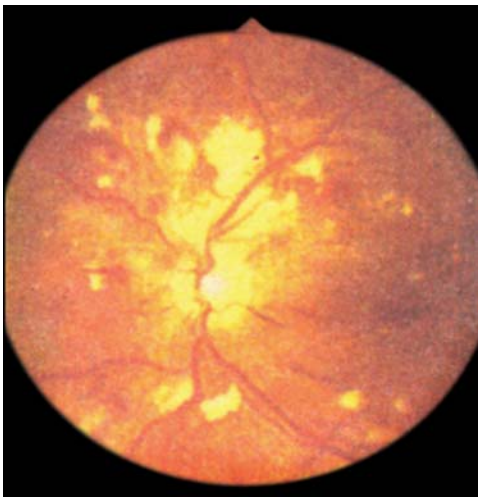


Fig. 18.23: Renal retinopathy
(*Courtesy*: Mr S Kanagami, Tokyo)

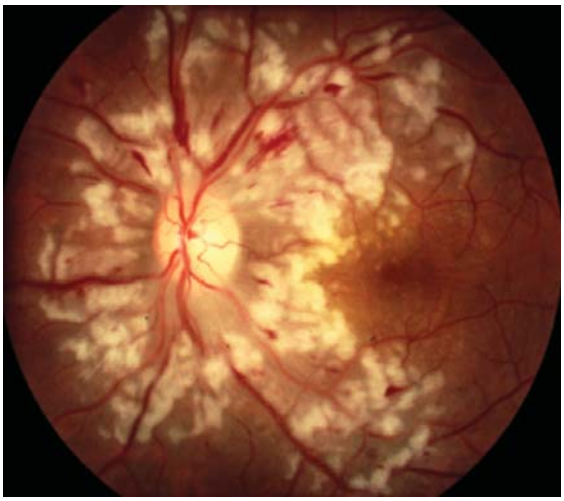


Fig. 18.24: Toxemia of pregnancy
(*Courtesy*: Mr S Kanagami, Tokyo)

retinopathy show degenerative changes in the form of hyaline or lipid degeneration.

Retinopathy in Toxemia of Pregnancy

Toxemia of pregnancy occurs in the later months of pregnancy (6-9 months) and is always accompanied with hypertension. In many respects the retinopathy is similar to hypertensive retinopathy (Fig. 18.24). The narrowing appears first in the nasal branches of the retinal arteries. Later, retinal edema supervenes and the picture resembles that of renal retinopathy. The retinal edema may be so marked that a bilateral exudative retinal detachment may develop causing profound loss of vision. The presence of retinopathy in toxemia of pregnancy warrants termination of pregnancy, since its continuance may result in the loss of vision and endanger the life of the mother as well as the fetus. Timely abortion leads to quick visual recovery.

Lupus Erythematosus Retinopathy

Systemic lupus erythematosus is a multisystem autoimmune inflammatory disease. It predominantly affects young women (female to male ratio is 9:1). Retinopathy develops in about 10 percent of the patients. Cotton-wool spots, flame-shaped hemorrhages (Fig. 18.25), microaneurysms and edema of the optic disk (papilledema) may be present.

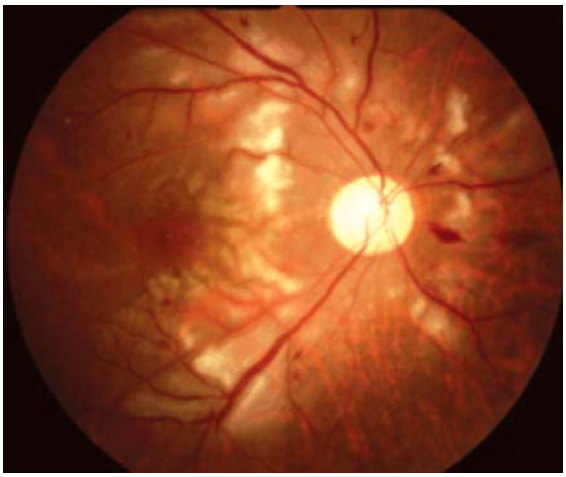


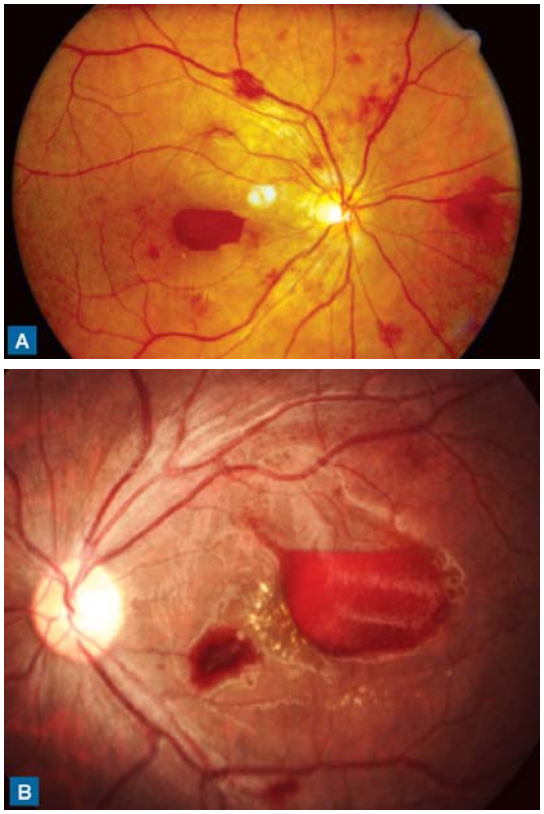
Fig.18.25: Lupus erythematosus retinopathy
(*Courtesy*: Mr S Kanagami, Tokyo)

Retinal Changes in Blood Dyscrasia

Retinal hemorrhages in the diseases of blood are common and perhaps occur as a result of deficient oxygenation leading to increased capillary permeability.

Retinopathy in Anemia

In severe anemia, the general fundus is pale and veins are dilated and multiple hemorrhages may occur (Fig. 18.26A). *White-centered hemorrhages*, a core of leukocytes surrounded by erythrocytes, and subhyaloid hemorrhage (Fig. 18.26B) are not uncommon at the posterior pole.



Figs 18.26A and B: Retinopathy in anemia: A. Multiple hemorrhages, B. Subhyaloid hemorrhage

Sickle-cell Retinopathy

Retinopathy may be found in patients with sickle-cell hemoglobin. The abnormal hemoglobin causes the red blood cells to assume a characteristic sickle-shaped appearance under hypoxic conditions. The sickle-cells can cause peripheral arteriolar occlusion in the retina.

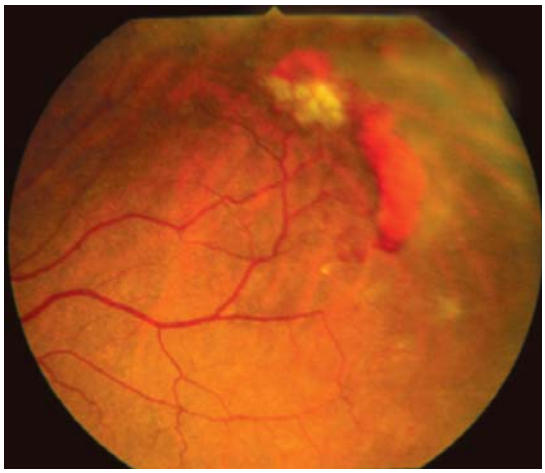


Fig.18.27: Sickle-cell retinopathy (*Courtesy*: Mr S Kanagami, Tokyo)

Proliferative retinopathy may develop in sickle-cell disease (Fig. 18.27). It is characterized by peripheral arteriolar occlusion, peripheral arteriovenous anastomosis, 'sea-fan' neovascularization (sprouting of new vessels in a fan-shaped manner), vitreous hemorrhage, vitreous traction and retinal detachment. Occasionally, a nonproliferative retinopathy may appear. It is marked by venous tortuosity, black sunbursts (peripheral chorioretinal scars), peripheral pink superficial hemorrhages, angioid streaks, sheathing of vessels and retinal breaks. Occlusion of the central retinal artery or vein is not rare.

Photocoagulation of the feeding arterioles and new vessels is the choice of therapy. Advance cases need bimanual vitrectomy.

Retinopathy in Leukemia

The fundus is pale and orange colored in leukemia. It shows characteristic white-centered hemorrhages scattered mostly in the peripheral retina. The retinal veins are dilated and tortuous. They appear bright red, while the arteries look pale yellowish-red and slightly constricted. Occasionally, leukemic deposits in the retina may be found.

Hemorrhages in the retina are common in purpura and polycythemia vera. Owing to plasma hyperviscosity, veins are enormously dilated and tortuous in polycythemia vera. It may also cause venous thrombosis and papilledema.

INFLAMMATORY DISEASES OF THE RETINA

Retinitis

Inflammation of the retina is called *retinitis*. It is often secondary to the inflammation of the choroid (chorioretinitis).

Primary inflammation of the retina is uncommon and may be classified as acute, subacute and chronic.

Acute purulent retinitis occurs due to the infection of retina by pyogenic organisms during septicemia, and may either lead to endophthalmitis or panophthalmitis.

Subacute infective retinitis or *septic retinitis of Roth* is due to the lodgement of septic emboli in the retina from bacterial endocarditis or from puerperal sepsis. It is characterized by the presence of round or oval white-centered hemorrhages (*Roth's spots*) at the posterior pole associated with retinal edema or papilledema.

Chronic granulomatous retinitis is usually secondary to choroiditis. However, *Treponema pallidum*, *Toxoplasma gondii* and cytomegalovirus can involve the retina primarily. These diseases have already been described in the chapter on *Diseases of the Uveal Tract*, but retinal lesions of syphilis need further elaboration.

Syphilitic Retinitis

Retina may be primarily involved in congenital as well as acquired syphilis.

Congenital syphilis causes anterior retinitis. It is marked by the presence of numerous black and white spots in the anterior retina giving a characteristic pepper and salt appearance. Large atrophic pigmented spots may also be found in the periphery of the retina.

Acquired syphilis causes a diffuse retinitis at the macula associated with exudates along the course of blood vessels. Yellow placoid macular lesions are characteristic of acute syphilitic chorioretinits. The chronic syphilitic retinitis often presents with depigmented retinal lesions, pigment aggregations, marked narrowing of the vessels and atrophy of the optic disk mimicking the fundus appearance in retinitis pigmentosa. The disease causes serious visual impairment and marked constriction of visual fields.

Retinal Lesions in AIDS

Acquired immune deficiency syndrome (AIDS) is characterized by a decrease in the CD4 subset of T-lymphocytes, an increase in the incidence of multiple opportunistic infections and progressive paralysis of the immune system of the body. It is caused by human immunodeficiency virus (HIV), a retovirus that relies on the enzyme reverse transcriptase for activity.

The retina is frequently involved in AIDS, about 58% of the patients show either vascular or infectious lesions. Ophthalmoscopically, discrete, fluffy opacities in the region of posterior pole adjacent to the major vascular arcades are seen in HIV retinopathy. They are cotton-wool spots (Fig. 18.28) appearing as a result of focal infarction of the nerve fiber layer. Flame-shaped hemorrhages, blot hemorrhages and microaneurysms are also seen.

Cytomegalovirus (*CMV*) *retinitis* is the most common ocular opportunistic infection in AIDS. It occurs when the CD4 T-cell count falls below 50. Patients complain of diminution of vision and black spots in their visual fields. The characteristic

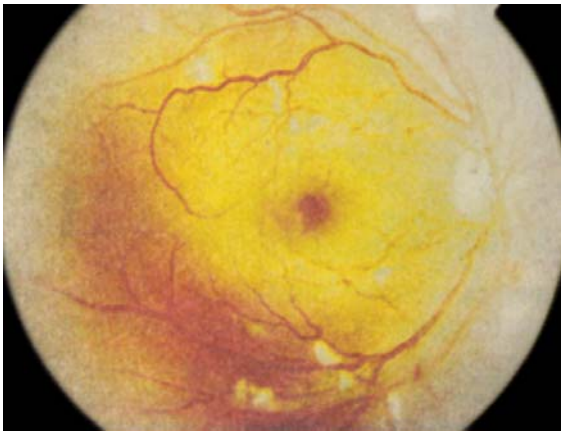


Fig. 18.28: Cotton-wool spots in HIV retinopathy

fundus picture of CMV retinitis includes intraretinal hemorrhages, exudates and retinal necrosis. The retina has an appearance of superficial granularity. Periphlebitis and vitreous inflammation may be seen.

Herpetic retinitis presents as progressive outer retinal necrosis (PORN) in AIDS that may be difficult to differentiate from peripheral CMV retinitis in the initial stages. However, rapid progression in a circumferential fashion and sparing of retinal vessels are typical of PORN. Bilateral involvement is the rule, although only one eye may be affected initially. PORN is characterized by early macular retinitis in the presence of little or no intraocular inflammation. Peripheral retina shows large areas of retinal whitening with outer retinal necrosis. Optic neuritis and vascular occlusion may be seen. Retinal breaks in necrotic retina may develop leading to rhegmatogenous retinal detachment.

Opportunistic syphilitic, mycobacterial, fungal or protozoal (toxoplasmosis) infections of the retina may also be observed in patients of AIDS.

HIV retinopathy is a noninfectious microvascular disorder and the retinal changes are not vision threatening. The advent of highly active antiretroviral therapy (HAART) has brought enormous changes in the scenario of AIDS. The prevalence of HIV retinopathy has diminished drastically due to early initiation of HAART.

Treatment of CMV retinitis includes 5 mg/kg twice daily of ganciclovir or 90 mg/kg twice daily of foscarnet for 2 weeks. After this high induction dose, the dose of the drug may be reduced depending on the patient's response to the treatment. Alternatively, cidofovir may be given intravenously once a week for 2 induction doses and then biweekly for maintenance. In non-responding cases intravitreal fomiversen, ganciclovir or foscarnet can be used. Intravitreal ganciclovir implant is also available.

Treatment of PORN includes systemic or intravitreal administration of ganciclovir. Prophylactic laser demarcation of the borders of necrosis reduces the risk of retinal detachment. Reattachment of retina is achieved by bimanual vitrectomy with silicone oil injection.

Eales' Disease

Eales' disease or periphlebitis retinae is a nonspecific inflammation of the veins of the peripheral retina.

Etiology

Eales' disease is not an uncommon disease and affects apparently healthy young adults, usually males. The etiology of the disease is unknown but tuberculosis and septic lesions anywhere in the body are implicated in its causation.

Clinical Features

The patient suffers from sudden loss of vision due to recurrent hemorrhages in the vitreous. Often the left eye is first to be affected, however, the hemorrhage may occur in the other eye within a

period of few weeks to months. The clinical manifestations of Eales' disease largely depend upon the extent of retinal vasculitis and obliteration of the affected vessels, especially the capillaries. The hypoxic retina produces a vasoproliferative substance causing neovascularization (Fig. 18.29). Initially, sheathing of small retinal veins accompanied with minute retinal hemorrhages may be found in the periphery of retina. Later, vitreous haze and peripheral retinal neovascularization develop. Fluorescein angiography reveals shunt vessels, staining of inflamed vessel wall, areas of capillary drop-out or retinal neovascularization. Recurrent vitreous hemorrhages can lead to massive retinitis proliferans (Fig. 18.30) and tractional retinal detachment. Rubeosis iridis, complicated cataract and neovascular glaucoma may occur in some eyes.

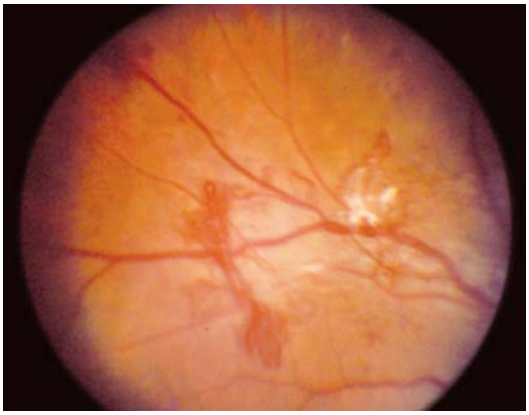


Fig. 18.29: Eales' disease: sheathing of veins and neovascularization

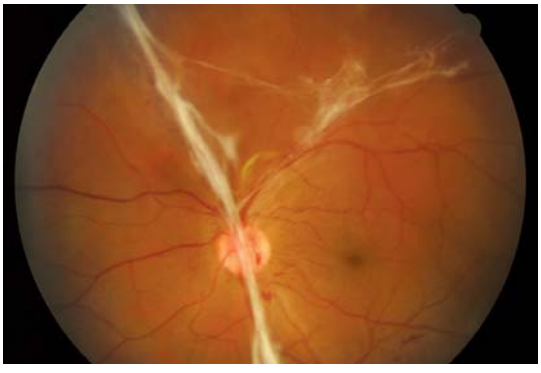


Fig. 18.30: Eales' disease: retinitis proliferans

Treatment

In the absence of proper etiology, the treatment of periphlebitis retinae is unsatisfactory. Treatment of tuberculosis and septic focus rarely prevents the recurrence of vitreous hemorrhage. Local and systemic corticosteroids have been used to control the intraocular inflammation. Photocoagulation of the neovascularized area has given an encouraging result (Fig. 18.31). Long-standing vitreous hemorrhage needs pars plana vitrectomy.

Central Serous Choroidopathy

Etiology

Central serous choroidopathy, also known as *central serous retinopathy* (CSR), is a central serous detachment of the neurosensory retina occurring in young males due to a defect in the pumping function of retinal pigment epithelium (RPE) associated with leakage of fluid from the choriocapillaris into the subretinal space. Type A personality, psychiatric drugs, stress and elevated blood levels of steroids have been implicated in the etiology of CSR.

Clinical Features

Blurring of vision, distortion of objects and seeing a black shadow before the eye are common symptoms. The macular area looks edematous

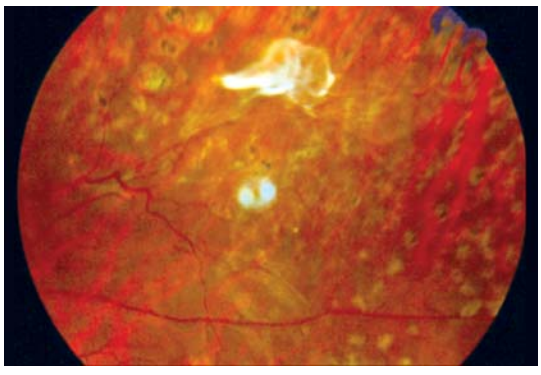


Fig. 18.31: Photocoagulation in Eales' disease

with loss of foveal reflex (Fig. 18.32A). A shallow localized detachment of the sensory retina at the posterior pole is seen with indirect ophthalmoscope. The lesion is more or less round with well-defined glistening borders.

Fluorescein angiography provides a definitive diagnosis of central serous retinopathy. In the beginning, a small hyperfluorescent spot (Fig. 18.32B) appears. It may expand in size and intensity as the angiogram progresses (*expansile dot*). Occasionally the dye may reach the subretinal space and ascend vertically in a *smoke-stack* manner (Fig. 18.32C) from the point of leakage to the upper limit of the detachment. Then it spreads laterally forming a *mushroom* or *umbrella pattern*.

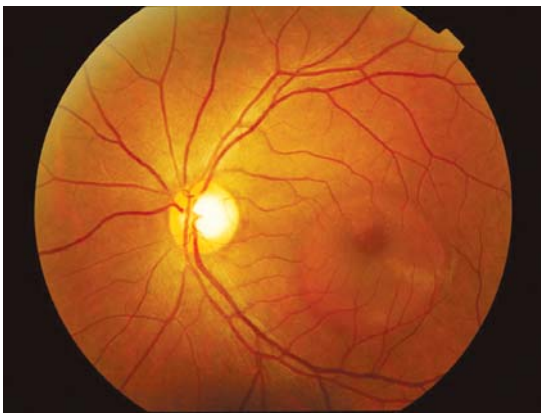


Fig. 18.32A: Central serous choroidopathy

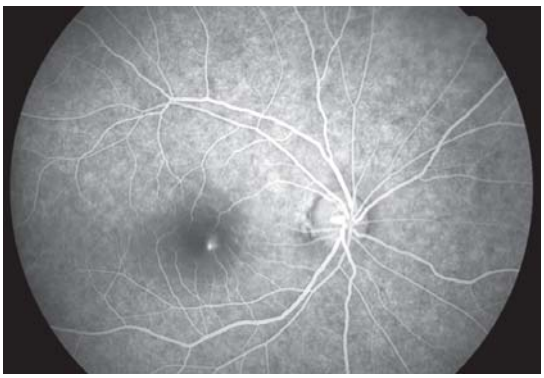


Fig. 18.32B: Central serous choroidopathy: FA showing hyperfluorescent spot

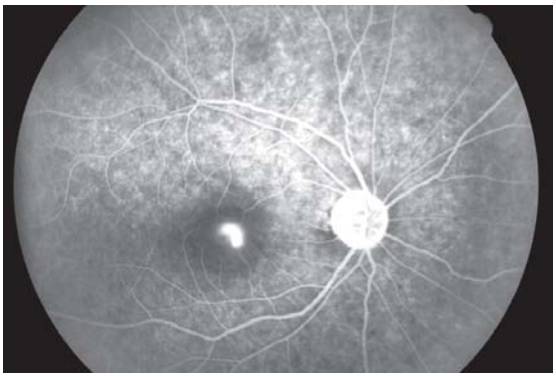


Fig. 18.32C: Central serous choroidopathy: FA showing smoke-stack pattern

The fluorescein angiography findings suggest that CSR is caused by a breakdown of blood-retinal barrier and, perhaps, a small defect in RPE.

Treatment

The condition is often transient and tends to resolve leaving minute yellow deposits in the deeper layers. When it persists for more than three to four months, secondary cystic changes in macula ensue. Permanent visual damage is likely to develop if there are repeated episodes of central serous retinopathy. However, the patients with non-involvement of fovea and detachment less than one disk diameter in size, carry good prognosis. Laser photocoagulation is indicated in patients with recurrent attacks or if the disease persists for 4 months or longer. The laser burns are applied to ablate the defective RPE, if it is not situated too near to the fovea, and the gap is bridged by the adjacent normal RPE cells.

Photoretinitis

Retinal burn may develop after gazing at a solar eclipse directly or indirectly (*solar retinopathy*) or accidental occupational exposure to arc welding without protective glasses. The damage occurs because of visible light or shorter wavelength ultraviolet radiation that causes photochemical

retinal injury. Young persons with clear crystalline lens are at a higher risk of developing retinopathy while patients with high refractive errors are less vulnerable. A persistence of after image phenomenon occurs soon after the exposure which causes decreased vision, positive scotoma, headache and metamorphopsia. On examination of the retina, initially, one may see a small yellow punctate spot which gets replaced by a red dot surrounded by a pigmented halo. After a couple of weeks a lamellar macular hole develops. In most of the patients the vision returns to the level of 6/6 to 6/9 within 3 to 6 months. Prevention by education and protection by smoked lenses or protective goggles is recommended. Once macula is charred no effective treatment is available.

DRUG-INDUCED MACULOPATHY

There are certain drugs that have a high affinity for melanin-containing structures of the eye. They get concentrated in the retinal pigment epithelium and the choroid causing maculopathy. Chloroquine, hydroxychloroquine, phenothiazines, tamoxifen and methoxyflurane may cause drug-induced maculopathies.

Chloroquine Maculopathy

Chloroquine is an antimalarial drug that is also used in the treatment of rheumatoid arthritis and systemic lupus erythematosus. The use of hydroxychloroquine is relatively safer than chloroquine. The long-term use of chloroquine causes keratopathy, maculopathy and optic neuropathy.

The corneal deposits (verticillata) do not cause visual symptoms but macular edema and subsequent development of maculopathy impair the vision significantly. The color vision is also affected. The characteristic lesion is a bilateral *bull's-eye maculopathy* (Fig. 18.33) with a central area of hyperpigmentation surrounded by a zone of hypopigmentation which in turn is surrounded by a region of hyperpigmentation.

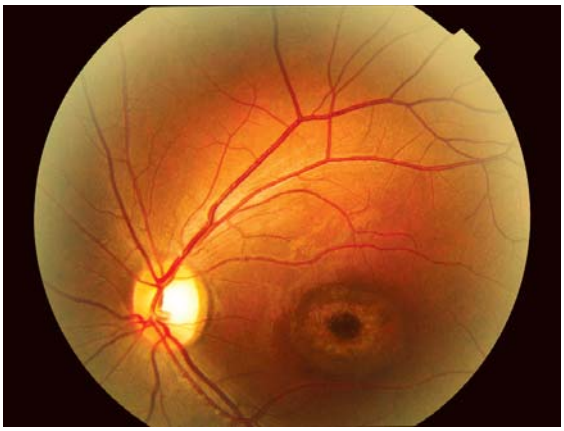


Fig.18.33: Bull's-eye maculopathy
(*Courtesy*: Dr Sanjay Thakur, Nataraj Eye Centre, Varanasi)

The visual fields testing often demonstrates bilateral paracentral scotomas, and FA reveals atrophy of the retinal pigment epithelium (transmission defect).

The drug should be immediately stopped in order to prevent the progress of maculopathy. The risk of maculopathy is low when recommended 3 mg chloroquine and 6.5 mg hydroxychloroquine per kg body weight are administered. Periodic eye screening is indicated in patients who are on long-term chloroquine.

DEGENERATIVE DISEASES OF THE RETINA

Retina undergoes degeneration either due to sclerosis of blood vessels or due to abiotrophy. Angiographic and histopathological studies have greatly contributed to the understanding of retinal degeneration.

Age-related Macular Degeneration (*Senile Macular Degeneration*)

Etiology

Age-related macular degeneration (AMD) is a major cause of blindness in persons of 50 years

and older. Besides advanced age, family history, female gender, ocular pigmentation, hypermetropia, hypertension, hypercholesterolemia, exposure to sunlight, cigarette smoking, malnutrition and cardiovascular disease are considered as risk factors.

AMD is classified into two types, *non-neovascular* or *atrophic* or *dry* and *neovascular* or *exudative* or *wet*. The patient often complains of blurring of vision, central black spot and distortion of image of an object.

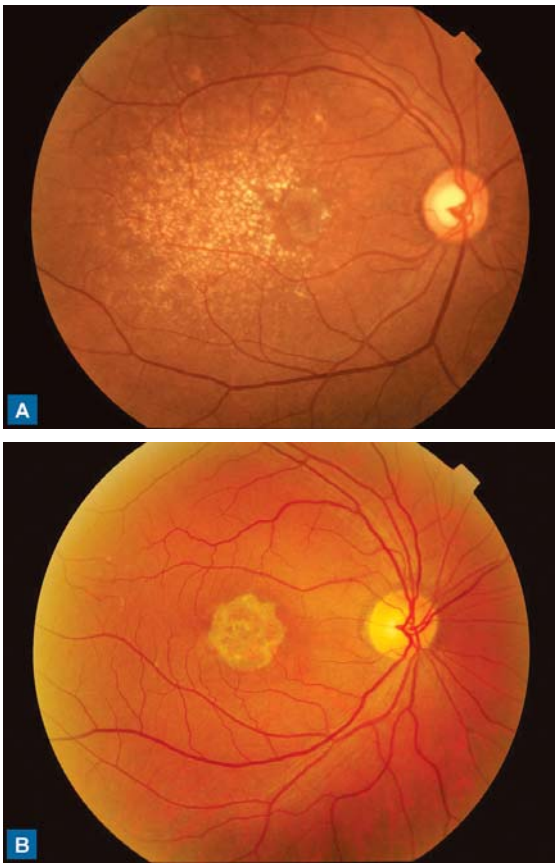
Non-neovascular AMD

In non-neovascular AMD multiple drusen, pigment clumping and areas of pigment atrophy (*geographic atrophy*) are found at the posterior pole (Figs 18.34A and B). Drusen are clinically seen as tiny, yellowish round lesions in the macular area at the level of outer retina. Histopathologically, they represent a thickening of inner portion of Bruch's membrane. The earliest morphologic feature of AMD is development of two distinct types of basal deposits beneath the RPE. The deposition of lipid-rich material with wide spacing of collagen fibers (*basal laminar deposits*) occurs between the plasma membrane and the basal lamina of RPE and accumulation of phospholipid vesicles and electron-dense granules (*basal linear deposits*) in the inner aspect of Bruch's membrane.

Drusen may be classified according to their sizes as *small* (less than 64 microns in diameter), *intermediate* (64 to 124 microns in diameter) and *large* (more than 124 microns in diameter). Intermediate and large drusen are pathognomonic of dry AMD.

Drusen may be *hard* with distinct borders, *soft* with poorly demarcated margins or *confluent*, contiguous with other drusen. Soft and confluent drusen may lead to geographic atrophy or choroidal neovascularization.

Patients with drusen may have normal or marginally diminished vision, with loss of



Figs 18.34A and B: Non-neovascular AMD

overlying photoreceptors. Fluorescein angiography shows staining of drusen in late phase of angiogram or pooling of dye in areas of diffuse thickening.

Non-neovascular AMD may cause atrophy of retinal pigment epithelial cells. Confluent areas of RPE atrophy making the choroidal vessels more readily visible characterize *geographic atrophy*. The photoreceptors lying over the area of geographic atrophy are usually attenuated. Fluorescein angiography shows typical window defect.

Focal hyperpigmentation at the level of outer retina shows areas of blocked fluorescence on angiography.

Treatment

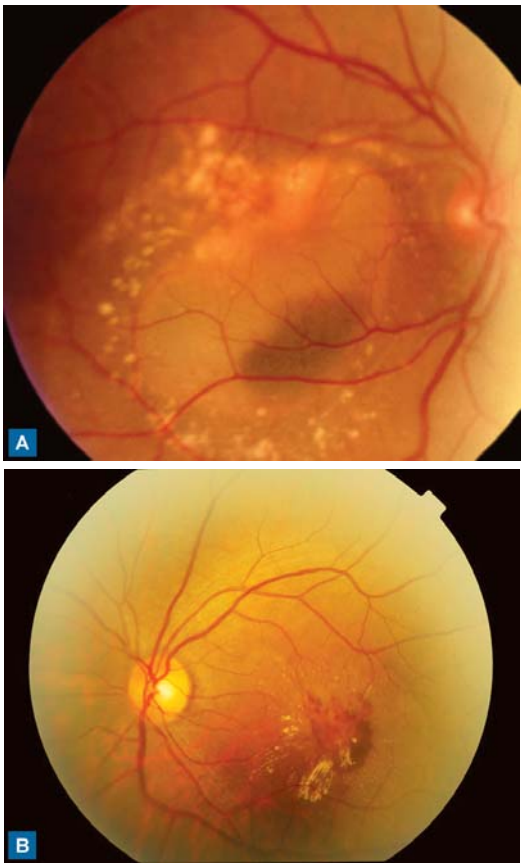
Patients with widespread intermediate drusen, large drusen, geographic atrophy and advanced AMD in one eye comprise high risk non-neovascular AMD. They should be given a combination of antioxidant vitamins (500 mg vitamin C, 400 IU vitamin E and 15 mg beta carotene) and zinc (80 mg zinc oxide and 2 mg cupric oxide to prevent zinc induced anemia) supplementation to decrease disease progression and visual loss according to the Age-Related Eye Disease Study (AREDS). Patients of non-neovascular AMD must be encouraged to quit smoking and to use sunglasses outdoors. They are trained to recognize symptoms of progression of the disease and ophthalmologists must look for signs of advanced AMD on every follow-up visit.

Neovascular AMD

Subretinal exudates and hemorrhages (Figs 18.35A and B), retinal pigment epithelial detachment (PED), choroidal neovascularization (CNV) and disciform scars are found in neovascular AMD. CNV is the hallmark of neovascular AMD. The non-neovascular changes of AMD lead to a break in Bruch's membrane through which fibrovascular complex from choriocapillaris proliferates within the inner aspect of Bruch's membrane. This fibrovascular tissue disrupts the normal anatomy of choriocapillaris, Bruch's membrane, RPE and photoreceptors.

Patients with wet AMD experience sudden diminution of vision, distortion of objects and positive scotoma. Slit-lamp biomicroscopy using 90 D lens shows subretinal exudates and hemorrhages, PEDs, pigment epithelial tears and a grayish neovascular membrane.

Fluorescein angiography reveals 2 patterns of CNV – classic and occult. *Classic CNV* shows a uniform hyperfluorescence in the early phase of angiogram which increases in size and intensity in the late views making the margin of lesion fuzzy. *Occult CNV* may be due to the presence of a fibrovascular PED which shows stippled hyperfluorescence in the early films of angiogram. The hyperfluorescence becomes intense in subsequent phases of angiogram but the stippled appearance persists.



Figs 18.35A and B: Neovascular AMD
(*Courtesy:* Mrs Kanagami, Tokyo)

Treatment

The mainstay of treatment for CNV is laser therapy. Photocoagulation is indicated for extrafoveal and juxtafoveal lesions with distinct boundaries. Laser treatment reduces the risk of progressive severe visual loss.

Photodynamic therapy (PDT) is indicated for subfoveal CNV, lesions too close to the foveal center, large sized lesions or CNV with poorly demarcated borders. A photosensitizing drug, verteporfin, is injected intravenously followed by application of diode laser. Exposure to laser light of 689 nm wavelength incites a photochemical reaction in the neovascular tissue, where the dye gets accumulated in high concentration, generating reactive oxygen species that leads to capillary endothelial cell damage and vessel thrombosis.

An anti-vascular endothelial growth factor (anti-VEGF) agent has recently been approved by FDA for use in patients with exudative AMD. Transpupillary thermotherapy (TTT), using 810 nm infrared diode laser, radiotherapy, submacular surgery to remove blood and neovascular tissue, and macular translocation have been tried in patients with neovascular AMD with variable success.

Angioid Streaks

Linear breaks in Bruch's membrane are called *angioid streaks*. They appear as brown irregular lines radiating from the disk and may be mistaken for blood vessels. They are situated deeper to the retinal vessels and are irregular in distribution. They are found in pseudoxanthoma elasticum, Ehlers-Danlos syndrome, sickle-cell anemia and Paget's disease.

Peripheral Retinal Degeneration

An assortment of degenerative lesions may be found in the peripheral retina. Some of the lesions are benign and do not predispose to the retinal breaks. Others, most often, lead to retinal breaks and retinal detachment.

Microcystoid degeneration, white-with-pressure and islands of dark brown pigmented areas in the periphery of retina do not predispose to retinal breaks. The presence of pavingstone degeneration and meridional folds at ora serrata have dubious role. Nevertheless, lattice degeneration and snail-track degeneration (Fig.18.36) can lead to retinal breaks and retinal detachment.

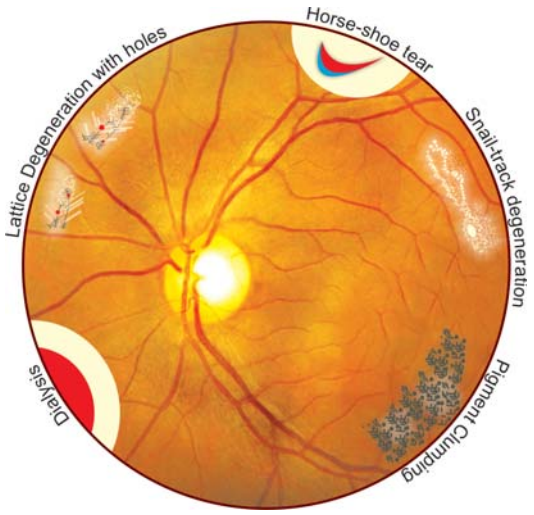


Fig. 18.36: Retinal degenerations and breaks

Lattice Degeneration

Lattice degeneration is the most significant retinal degeneration as it often leads to retinal breaks and retinal detachment. A classical lattice degeneration consists of well-demarcated, circumferentially oriented, somewhat spindle-shaped areas of retinal thinning, commonly found between the equator and the ora. It starts as an area of fine white stippling or white shining dots that gives a frost-like appearance on scleral depression during indirect ophthalmoscopy. The traction on the lattice by vitreous may result in horse-shoe tears and subsequent retinal detachment. Prophylactic laser demarcation or transconjunctival cryopexy of the lattice is advocated.

Snail-track Degeneration

Snail-track degeneration resembles closely with lattice degeneration. It consists of well-demarcated linear areas of white dots or snow-flakes that give a frost-like appearance.

Degenerative Retinoschisis

Degenerative retinoschisis is an acquired splitting of the layers of peripheral retina. The condition is

usually bilateral, symmetrical and often asymptomatic. Retinoschisis should be treated when it threatens the macula. A barrage laser photocoagulation is used, or cryopexy around the holes may be performed.

Primary Pigmentary Retinal Degeneration

A bilateral progressive loss of vision beginning with night-blindness and associated with bone corpuscular pigment deposits, narrowed arteries and optic atrophy characterize primary pigmentary retinal degeneration often referred as *retinitis pigmentosa* (RP).

Etiology

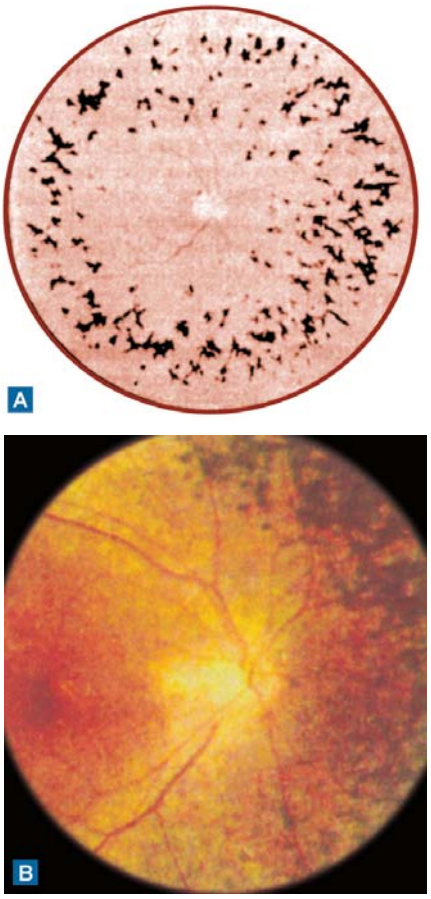
The condition is inherited as a recessive trait with a 20 percent incidence of consanguinity of the parents. Occasionally, it shows a dominant inheritance. The sex-linked variety is also documented which is more severe than the others.

The severity of disease varies with the site of mutation in the rhodopsin gene. Rhodopsin, a visual pigment found in the rods, is responsible for night vision. Mutations in periferin/retinal degeneration slow (RDS) gene have wide disease expression. RDS gene encodes periferin, a glycoprotein present in the peripheral portion of photoreceptors.

There may be a large number of patients of RP with no family history of the disease (*simplex RP*). The exact etiology of retinitis pigmentosa (RP) is, therefore, unknown. It is considered as an abiotrophy and primarily affects rods and cones, particularly the former, and the retinal pigment epithelium. The degeneration starts in the equatorial zone and slowly spreads both anteriorly and posteriorly.

Clinical Features

Defective vision in twilight or night-blindness is the most prominent symptom of the disease. Later, progressive contraction of the visual field handicaps the patient even in moving around. Ophthalmoscopic examination may not reveal any sign initially. A small irregular pigment mottling is found in the equatorial zone, from here the pigmentary changes extend both towards the posterior pole and the ora serrata. As the disease progresses, characteristic small jet-black pigments resembling bone spicules with spidery outlines appear in the entire retina especially along the course of the retinal veins (Figs 18.37A and B). These pigments lie anteriorly thereby they hide the course of vessels. In contrast, the choroidal atrophic spots lie in a deeper plane and the retinal vessels course over them. With the anterior migration of pigments from the retinal pigment epithelium (RPE) the choroidal vessels are often visible and fundus becomes tessellated.



Figs 18.37A and B: Typical retinitis pigmentosa

The retinal arterioles are affected early and become extremely attenuated. Similar changes may follow in veins also. The optic disk becomes pale waxy and atrophic—*consecutive optic atrophy*.

Macula is often involved in retinitis pigmentosa. A cystoid maculopathy may be found in about 70% of the eyes with retinitis pigmentosa, although the patients retain a relatively good visual acuity. Atrophic lesions of RPE are fairly common. Keratoconus, open-angle glaucoma, myopia and posterior vitreous detachment are the other associated ocular findings. Long-standing cases of retinitis pigmentosa develop complicated cataract.

In the early stage of retinitis pigmentosa, the characteristic pigments do not manifest and other fundus changes are minimal. However, the diagnosis can be made with the help of electroretinography (ERG). The ERG, especially the scotopic component, is markedly subnormal (Fig. 18.38). Later, it becomes extinguished. Electro-oculographic (EOG) change also develops early and shows an absence of light rise. The dark adaptation time is always increased.

The visual field defects in RP are quite characteristic. Initially, an incomplete or complete ring scotoma (Fig. 18.39) may be found corresponding to the degenerated zone near the equator. Gradually, the visual field shows concentric contraction, particularly marked when the illumination is reduced. Finally, a small area around the fixation point is retained and, thus, the patient has only tubular vision. Despite the tubular vision, the patient may retain good central visual acuity and be able to read and write. However, the vision is seriously affected after the age of 50 years due partly to advanced degenerative changes and partly to complicated cataract.

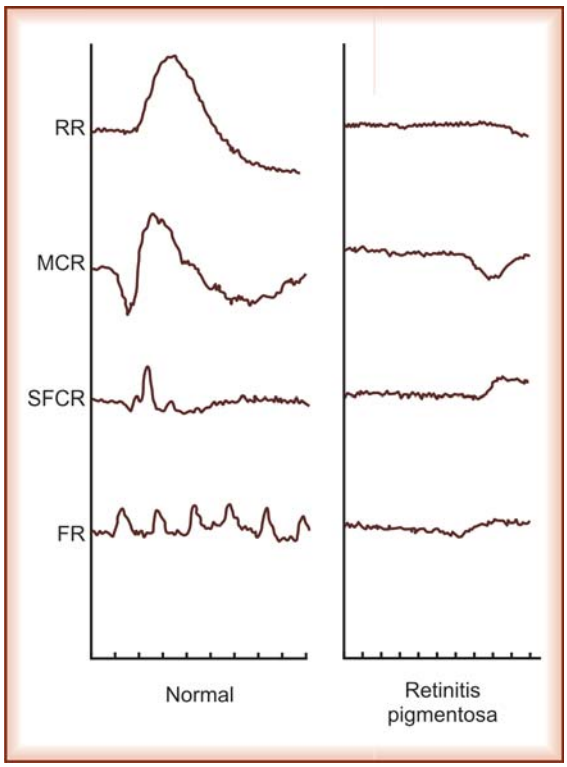


Fig. 18.38: ERG changes in RP

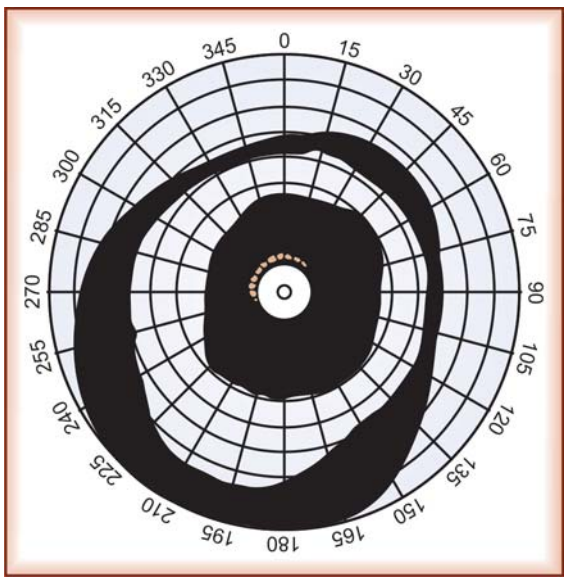


Fig.18.39: Complete ring scotoma in RP

Atypical Retinitis Pigmentosa

Retinitis pigmentosa shows great clinical variations. *Retinitis pigmentosa sine pigmento* is a variant

of retinitis pigmentosa in which the fundus does not have any visible pigment deposit. However, night-blindness, visual field defect and subnormal ERG clinch the diagnosis.

Retinitis punctata albescens is yet another variety of retinitis pigmentosa characterized by the presence of numerous small white dots distributed all over the fundus and associated with typical symptoms of the disease.

Inverse retinitis pigmentosa is characterized by the lesion confined to the macular region and field loss progressing outward from the centre (*central RP*). Sporadic cases of RP may present with a ring scotoma within the central 20-30 degrees—*pericental RP*.

When 1 or 2 sectors of the retina are involved the condition is known as *sectorial RP*. The disease is bilateral and slowly progressive. It is believed to be due to ultraviolet light damage to the retina. Dark goggles and antioxidants are recommended for patients with sectorial RP.

Unilateral RP, wherein only one eye is affected, may occur occasionally.

Differential Diagnosis

Common causes of acquired pigmentary retinal degeneration include trauma, inflammation (syphilis, rubella, Harada's disease), ophthalmic artery occlusion, spontaneously reattached retina and retained metallic intraocular foreign body. RP should be differentiated from other causes of pigmentary retinal degeneration by ERG and visual field testing.

Secondary Pigmentary Retinopathy

Retinitis pigmentosa may be associated with systemic diseases—*secondary pigmentary retinopathy*. *Bardet-Biedel syndrome* combines RP, obesity, mental retardation, polydactyly or syndactyly and hypogenitalism. *Laurence-Moon syndrome* is similar to Bardet-Biedel syndrome except that there occurs no obesity and polydactyly, but these patients develop spastic paraplegia. Association of congenital sensorineural hearing loss and RP is seen in *Usher syndrome*.

Refsum's syndrome is characterized by RP, hearing loss, cerebellar ataxia and polyneuropathy. It is a hereditary disorder of lipid metabolism wherein there occurs an absence of phytanic acid oxidase. As a result phytanic acid accumulates in the body tissues.Treatment with low phytol and low phytanic acid diet retards neurologic complications and retinal degeneration.

Abetalipoproteinemia is an autosomal recessive disease characterized by RP, acanthocytosis, fat malabsorption and fat soluble vitamin deficiency. Treatment with vitamin A and E prevents the retinal degeneration.

Treatment

There is no specific therapy for retinitis pigmentosa. Low vision aids may be tried in patients with subnormal vision. Advanced cases are advised vocational rehabilitation and mobility training. The advancement in molecular genetics in recent years has given some hope of replacing or regularizing the diseased gene. Clinical trials are going on to transplant retinal cells in order to find a remedy for this chronic degenerative disease.

The patients must be followed at 1-2 years interval. The progression of the disease is monitored by recording of visual field and electroretinography.

Most of the patients of RP retain at least some useful vision and total blindness is a rarity. They may not be disallowed to have children unless they are suffering from the autosomal dominant disease.

MACULAR DYSTROPHIES

Macular dystrophies are characterized by an early age of onset of diminution of vision and a slowly

progressive course. They are familial, bilateral and show a symmetrical involvement. They may involve the nerve fiber layer, photoreceptors or retinal pigment epithelium.

Juvenile Retinoschisis

Juvenile retinoschisis is a sex-linked recessive condition occurring virtually exclusively in hypermetropic male patients. It is caused by splitting of the retina at the level of nerve fiber layer. It is characterized by cystoid spaces with bicycle-wheel pattern of radial striae due to schitic changes within the foveola.

Cone Dystrophy

Cone dystrophy is seen between first and third decades of life with impairment of central vision, photoaversion or light intolerance, color blindness and nystagmus. A classical *bull's-eye maculopathy* is seen. ERG is markedly reduced especially the photopic component. Patients with cone dystrophy must be differentiated from other causes of bull's-eye maculopathy that include Stargardt's disease, chloroquine maculopathy, age-related macular degeneration (atrophic), central areolar choroidal dystrophy, longstanding macular hole and Batten disease.

Stargardt's Macular Dystrophy

Stargardt's macular dystrophy is an autosomal recessive condition characterized by an elliptical atrophic maculopathy giving a beaten-bronze appearance (Fig. 18.40). A broad ring of flecks usually surrounds this area. It leads to bilateral gradual diminution of vision during first or second decade of life. EOG is abnormal but photopic ERG tends to be normal.

Fundus Flavimaculatus

Fundus flavimaculatus and Stargardt's macular dystrophy are now regarded as variations of the same disorder. Fundus flavimaculatus is seen usually later than Stargardt's disease and characterized by round, oval or pisciform (fish-tail-like) yellow flecks scattered throughout the posterior poles of both eyes. It is generally detected on routine eye examination and the patient has a normal vision, but if fovea is involved, the vision gets impaired.

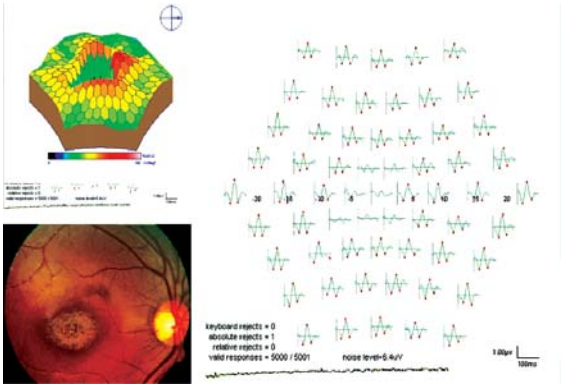


Fig. 18.40: Stargardt's disease
(*Courtesy:* Dr Subhadra Jalali, LVPEI, Hyderabad)

Familial Dominant Drusen

Familial dominant drusen is seen between second and third decades of life. Round, sharply defined yellow dots or flecks arranged in a mosaic or honeycomb pattern (*Doyne's honeycomb dystrophy* or *Tay's choroiditis*) are seen at the posterior pole of both eyes. The flecks are rounder, whiter and more sharply delineated than those in fundus flavimaculatus. Later they tend to become confluent and retina shows pigment epithelial atrophy. Initially the patients are symptom-free but later vision decreases.

Best's Vitelliform Dystrophy

Best's vitelliform dystrophy is an autosomal dominant condition occurring between first and second decades of life. There occurs a gradual diminution of vision over a period of years. The ophthalmoscopic features of Best's dystrophy are divided into five stages:

1. *Previtelliform stage*: Fundus is normal but EOG is abnormal.

2. *Vitelliform stage:* A yellowish round lesion (egg-yolk) is seen at the posterior pole.
3. *Pseudohypopyon stage*: A part of egg-yolk gets absorbed or disintegrated resulting in a cyst formation.
4. *Vitelliruptive stage*: It presents a scrambled egg appearance with visual impairment.
5. *End stage*: Macular scar or choroidal neovascularization develops with severe loss of vision.

NEW FORMATIONS IN THE RETINA

Phacomatoses

Phacomatoses form a group of familial diseases having a tendency for the development of neoplasm in the central nervous system, skin and eye. The conditions are transmitted as autosomal dominant traits. The phacomatoses comprise angiomatosis of the retina, tuberous sclerosis, neurofibromatosis and Sturge-Weber syndrome.

Angiomatosis retinae (*von Hippel disease*) is an uncommon condition occurring in men in the third and the fourth decade of life. It is characterized by marked tortuousity and dilatation of vessels, aneurysms and balloon-like capillary angioma in the retina (Fig. 18.41). Later massive exudation may cause retinal detachment. In 20 % of patients, the retinal lesions are associated with angiomatosis of cerebellum, medulla oblongata, spinal chord, kidney, adrenals and pancreas (*von Hippel-Lindau disease*). Early destruction of retinal angiomas by photocoagulation or cryotherapy has been found to be beneficial in the prevention of exudative retinal detachment.

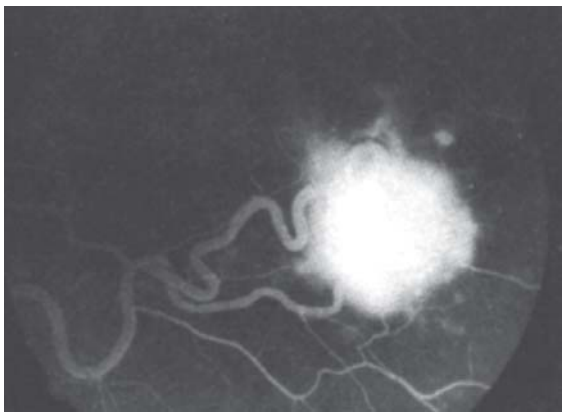


Fig. 18.41: Angioma of retina on FA

Tuberous sclerosis (Bourneville's disease) is characterized by nodular lesions on the face or *adenoma sebaceum* (Fig. 18.42), retinal mulberry tumors of the size of optic nerve head (Fig. 18.43) particularly near the optic disk, central nervous

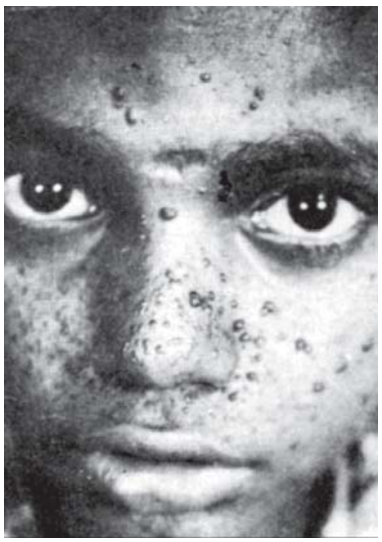


Fig. 18.42: Bourneville's disease with characteristic adenoma sebaceum

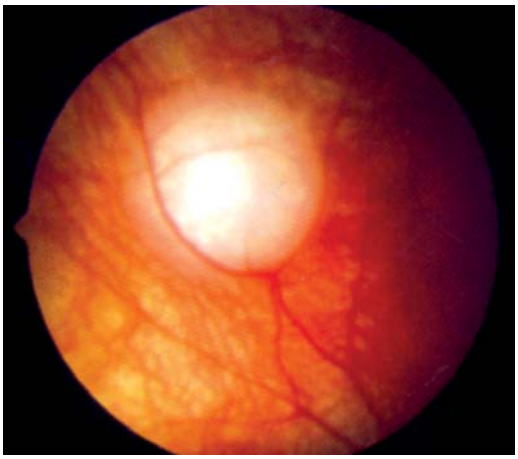


Fig. 18.43: Astrocytoma
(*Courtesy* Prof. DM Robertson, Rochester, MN)

system (CNS) tumors, renal tumors and multiple cysts in the lungs. It affects young individuals who suffer from convulsive seizures and mental retardation due to cerebral lesions. Treatment is of no avail and the prognosis is poor.

Neurofibromatosis (von Recklinghausen's disease) is a generalized hereditary disease charaterized by multiple small tumors of the skin (Fig. 18.44), peripheral nerves and CNS, *café-au-lait* spots and bone defects. The hypertrophied nerves can be palpated through the skin as hard cords or knobs. Neurofibromatosis of the lids may be associated with infantile glaucoma or intracranial gliomas. The disease can erode the orbital periosteum and may lead to pulsating exophthalmos.

Sturge-Weber syndrome is marked by the presence of capillary hemangioma or *nevus flammeus* on one side of the face (Fig. 18.45), hemangioma of the choroids, angioma of the meninges and infantile glaucoma on the affected side. The intracranial lesions cause Jacksonian epilepsy, and may undergo calcification which may be demonstrated on radiography.

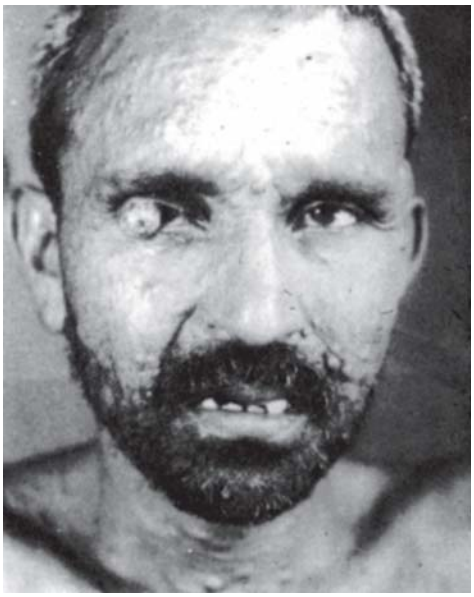


Fig. 18.44: von Recklinghausen's disease—neurofibroma of the right upper lid

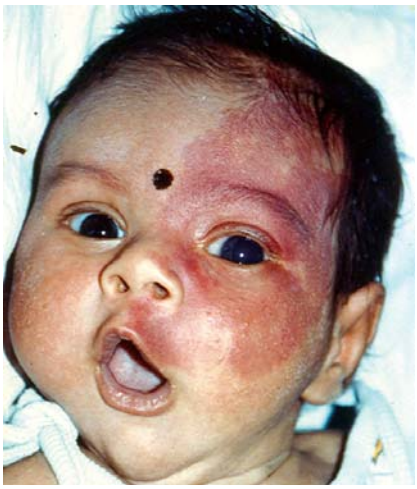


Fig.18.45: Sturge-Weber syndrome—facial hemangioma (*Courtesy*: AK Mandal LVP eye institute, Hyderabad)

DETACHMENT OF THE RETINA

Developmentally, there lies a potential space between the pigment epithelium and rest of the layers of the retina. In certain pathological conditions, the neurosenory retina is separated from the underlying pigment epithelium—such a condition is called *retinal detachment*. Strictly speaking, it is a misnomer and it represents only a separation of the retina. A true retinal detachment is that condition wherein the entire retina (including the pigment epithelium) is separated or pulled away from its bed, i.e., the choroid.

Retinal detachment (RD) can be clinically classified into two categories.

1. Rhegmatogenous or primary or simple RD associated with retinal break(s), and
2. Nonrhegmatogenous or secondary RD.

Rhegmatogenous RD

Pathogenesis

The detachment of retina associated with the formation of a break in the retina is known as *rhegmatogenous detachment* (Fig. 18.46). The rhegmatogenous RD almost always occurs due to formation of a break allowing the liquified

Fig. 18.46: Rhegmatogenous retinal detachment

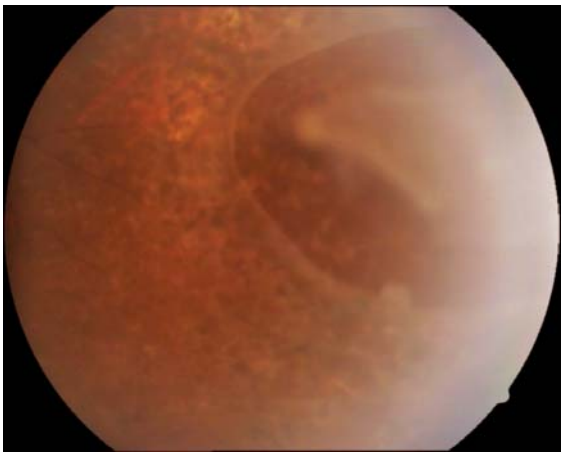


Fig. 18.47: Horse-shoe tear
(*Courtesy*: Dr YR Sharma, Dr RP Center, New Delhi)

vitreous to seep between the pigment epithelium and rest of the retina. It is bilateral in about 10% of cases. Retinal detachment is seldom found if the vitreous gel is healthy. But if the vitreous is fluid and adherent to the retina, it exerts a dynamic traction on the retina during the ocular movements and produces a *retinal tear* and subsequent detachment. Sometimes, an atrophic round break, *retinal hole*, is the precursor of an RD. A direct trauma may produce small or *giant retinal tears* (GRT).

Nearly 60 percent of breaks develop in the peripheral retina that shows degenerative changes. Age-related liquefaction of the vitreous gel (*synchysis senilis*) may predispose to horse-shoe tear (Fig. 18.47) due to transmission of traction at the site of vitreoretinal adhesion. Retinal holes often develop in atrophic retina and are commonly found in high myopia. About 40 percent of all retinal detachments occur in myopic eyes. Trauma, blunt or penetrating, is another important cause of RD. A blunt trauma may cause retinal dialysis or equatorial or macular hole. A penetrating trauma leads to the formation of a retinal break by the direct impact of a foreign body or by traction in approximately 20 percent of eyes. RD is more common in aphakic eyes (more common following intracapsular cataract extraction than extracapsular cataract surgery with or without intraocular lens implantation).

Clinical Features

The symptoms of retinal detachment are variable. Shallow detachment may not present much visual impairment as the retina still gets its nourishment from the underlying choriocapillaris. Transient flashes of light (photopsia) are the most common initial symptoms of the detachment which occur due to irritation of the neurosensory retina. The patient usually complains of distortion of objects and numerous black spots (floaters) in the field of vision. Later, the vision becomes foggy or cloudy due to extensive detachment.

The patient narrates that a veil has descended in front of the eye and the objects in the upper or lower visual fields are not visible. This can be confirmed by visual field charting; an absolute scotoma corresponds to the sector of the detachment of the retina. In spite of visual impairment, the central vision remains unaffected for sometime. When detachment progresses, it involves the macular region and affects the central vision as well. Rarely, the central vision is first to go if a macular hole develops.

In a case of RD, the anterior segment examination by slit-lamp reveals fine pigmented cells or tobacco-dust either on the anterior face of vitreous (*Shafer's sign*) or in the anterior chamber. With no past history of ocular trauma or intraocular surgery or inflammation, it is pathognomonic of a retinal break.

For the examination of the anteriormost part of the retina, the use of an indirect ophthalmoscope is indispensable. The shallow detachment of the retina presents a diagnostic riddle for the beginners as the color of the detached portion is not much different from that of the undetached retina. Gradually, the detached retina assumes a white or gray discoloration with folds or corrugations which may oscillate on ocular movements. The retinal vessels coursing over the detached retina look darker than usual. There may be more than one break that may remain hidden beneath the retinal folds. Repeated meticulous examinations after full mydriasis are necessary to discover such breaks. Most peripheral lesions require the use of a scleral depressor. Accurate localization of retinal breaks is essential. A careful preoperative drawing showing the position of retinal breaks and the extent of detached retina must be made.

A retinal detachment of recent origin tends to lower the intraocular pressure, but a retinal detachment of long-standing duration causes elevation of pressure. The elevated ocular tension is probably due to a low grade uveitis produced by the detachment, causing particulate material to obstruct the outflow channels. In the absence of uveitis, one may suspect that open-angle glaucoma preceded the retinal detachment (12-17 %). Long-standing retinal detachment leads to development of a demarcation line between the detached and attached retina (Fig. 18.48), retinal thinning and proliferative vitreoretinopathy (PVR). PVR results in vitreous haze, wrinkling of the retina, retinal stiffness, rolled edges of breaks and rigid retinal folds. Chronic RD leads to the formation of complicated cataract owing to disturbed metabolism of the lens.

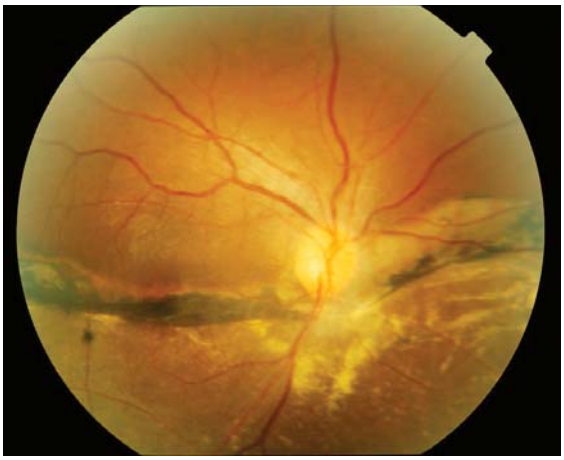


Fig. 18.48: Pigmented demarcation line between attached and detached retina (*Courtesy*: Dr Sanjay Thakur, Nataraj Eye Centre, Varanasi)

Treatment

The main objective of treatment of a retinal detachment is to seal and support the retinal break. The break can be sealed in three ways: cryopexy, photocoagulation and diathermy. A scleral buckle supports the break by reducing the dynamic vitreoretinal traction as well as apposing the RPE to the neurosensory retina (See video). Retinal detachment with GRT or PVR requires pars plana vitrectomy. Late and untreated cases have poor prognosis. Prophylactic photocoagulation or cryopexy is recommended in high myopic patients with lattice degeneration and retinal breaks.

Nonrhegmatogenous Retinal Detachment

Nonrhegmatogenous RD is divided into two categories:

1. Exudative RD, and
2. Tractional RD.

Table 18.1: Causes of exudative retinal detachment

Lesions	*Causes*
Transudative	Toxemia of pregnancy, renal hypertension, bullous central serous retinopathy
Exudative	Harada's disease, posterior scleritis or uveitis, orbital cellulitis
Neoplastic	Malignant melanoma, retinoblastoma (exophytum type)

Exudative Retinal Detachment

The absence of a break in the retina and shifting of subretinal fluid on changing the position of head are hallmarks of an exudative retinal detachment. Exudative RD is less common than rhegmatogenous RD. Inflammatory or neoplastic lesions are the leading causes of exudative detachment (Table 18.1).

The patient may complain of diminution of vision and black mobile spots before the eye. However, photopsia is absent. Funduscopy reveals a smooth retinal surface without corrugations. A shift in subretinal fluid with changing the position of head is suggestive of an exudative RD. Absence of retinal break or proliferative vitreoretinopathy helps in establishing the diagnosis of exudative detachment.

Treatment

Transudative, exudative or hemorrhagic RD may undergo spontaneous regression following absorption of the fluid. Ocular inflammatory diseases need speedy management to avoid visual loss. Neoplastic lesions require special attention.

Tractional Retinal Detachment

Tractional RD occurs due to contraction of the membrane in the vitreous that pulls away the sensory retina from the retinal pigment epithelium. Proliferative diabetic retinopathy and trauma are the most common causes of tractional RD. Other causes include Eales' disease, chronic uveitis and retinopathy of prematurity.

Recurrent bleeding in the vitreous acts as a stimulus for fibroblastic proliferation. The contraction of bands or epiretinal membrane (Fig. 18.49) over areas of strong adhesions detaches the retina. The retina is immobile with a smooth surface. The detached retina assumes a concave configuration towards the front of eye in the absence of a break. However, a small hole may develop posterior to the equator, the RD then assumes a convex bullous configuration. Besides RD, other features of the causative disease may be found.

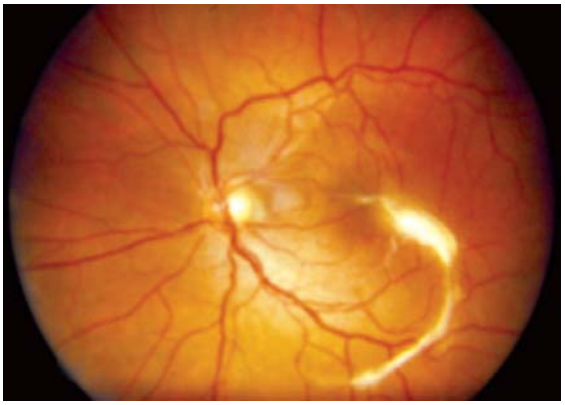


Fig. 18.49: Epiretinal membrane

Treatment

The tractional pull is relieved by segmentation or delamination. The epiretinal membrane can be removed by peeling during pars plana vitrectomy along with a prophylactic encirclage.

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CHAPTER

19 Diseases of the Optic Nerve

ANATOMY

The optic nerve is a part of the visual pathway. It is mainly composed of the nerve fibers derived from the ganglion cells of the retina terminating in the lateral geniculate nucleus. A small number of pupillomotor fibers and some centrifugal fibers are also present. The optic nerve contains about one million fibers. Initially, the macular fibers lie in the lateral part of the nerve but they assume a central position as the nerve passes backwards through a short circular scleral opening situated 1 mm above and 3 mm nasal to the posterior pole of the eye. The fibers from the peripheral parts of the retina enter the periphery of the optic nerve. A partial decussation occurs in the chiasma, wherein the nasal fibers cross while the temporal ones enter the optic tract of the same side to reach the lateral geniculate nucleus.

Optic Nerve

The optic nerve measures about 5 cm. It may be divided into four parts—intraocular, intraorbital, intracanalicular and intracranial.

1. *Intraocular part* of the optic nerve or optic nerve head (ONH) begins at the optic disk and extends to the posterior scleral surface. It measures approximately 0.7 mm and represents the confluence of 1-1.2 million axons of the ganglion cells.
2. *Intraorbital part* extends from the back of sclera to the orbital end of the optic foramen. It is undulant and measures about 3 cm in length. Posteriorly it is in closed proximity with the annulus of Zinn.
3. *Intracanalicular part* measures 6 mm. The ophthalmic artery crosses the nerve inferiorly in the dural sheath to lie on its lateral side. A thin bone separates the sphenoidal and ethmoidal sinuses medially from the nerve. Therefore, infection of these sinuses may cause optic neuritis.
4. *Intracranial part* of the optic nerve extends from the posterior end of the optic foramen to the anterolateral angle of the optic chiasma measuring about 1 cm. It lies above the cavernous sinus.

Sheath of the Optic Nerve

The optic nerve in the cranial cavity is surrounded by pia mater but arachnoid and dura are added to it in the intracanalicular part. The arachnoid terminates at the posterior part of lamina cribrosa by fusing with the sclera, while the dura mater becomes continuous with the outer two-thirds of the sclera.

Lamina Cribrosa

The lamina cribrosa is a sieve-like structure which bridges across the scleral canal. The lamina gets

its blood supply from the circle of Zinn. There is a funnel-shaped depression in the center of the optic nerve head which is called as *physiological cup*.

Blood Supply of Optic Nerve

The blood supply of the optic nerve resembles more or less that of the brain (Fig. 19.1). It is mainly through the pial network of vessels except in the orbital part which is also supplied by an axial system. The pial plexus is derived from the branches of ophthalmic artery, the long posterior ciliary arteries, the central retinal artery and the circle of Zinn. The circle of Zinn-Haller is an intrascleral peripapillary arteriolar anastomosis derived from short posterior ciliary arteries and supplies the intraocular part of optic nerve. The venous drainage of optic nerve occurs through the central retinal vein and pial plexus.

DISEASES OF THE OPTIC NERVE

The diseases of the optic nerve may be classified as congenital, circulatory, inflammatory, degenerative and neoplastic.

CONGENITAL ANOMALIES OF THE OPTIC NERVE

Coloboma of the Optic Disk

Typical coloboma of the optic disk (Fig. 19.2) occurs due to an incomplete closure of the optic fissure. It may be associated with an extensive coloboma of the fundus. It usually manifests as an inferior crescent resembling the myopic crescent to some extent. The eye is usually hypermetropic and astigmatic. The crescent is often ectatic or appears like a conus. The eye with colobomatous defect has a superior visual field defect and decreased vision. The coloboma of the optic disk may be confused with glaucomatous cupping.

Congenital Pit of the Optic Disk

A round or oval pit (Fig. 19.3) in the inferotemporal quadrant of the optic disk may be found. It appears darker than the usual color of the disk and is often associated with a serous detachment of the retina mimicking central serous retinopathy.

Hypoplasia of the Optic Nerve

Hypoplasia of optic nerve is a bilateral symmetrical condition characterized by a small disk, small tortuous vessels and peripapillary halo of hypopigmentation (double ring sign). Binasal or bitemporal visual field defects are common.

Morning Glory Syndrome

Morning glory syndrome is a unilateral condition of dysplastic coloboma of the optic nerve head

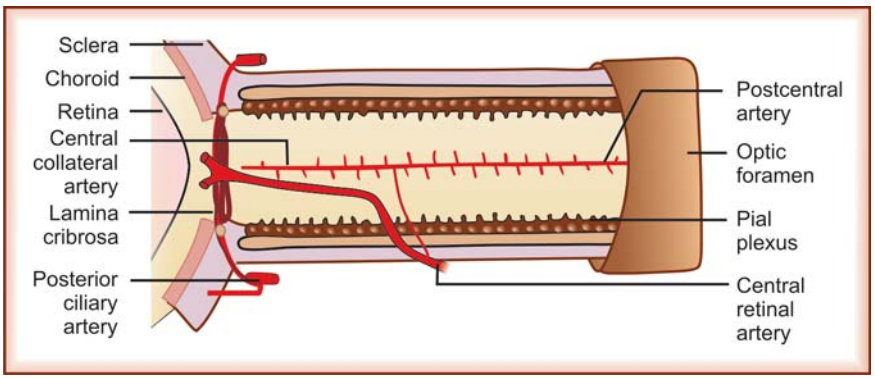

Fig. 19.1: Diagrammatic representation of blood supply of optic nerve

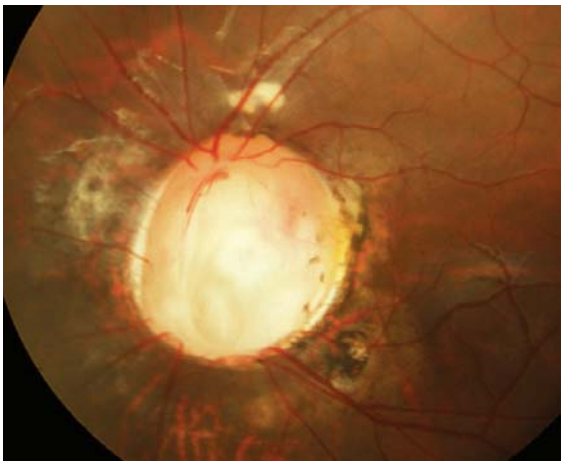

Fig. 19.2: Coloboma of optic disk

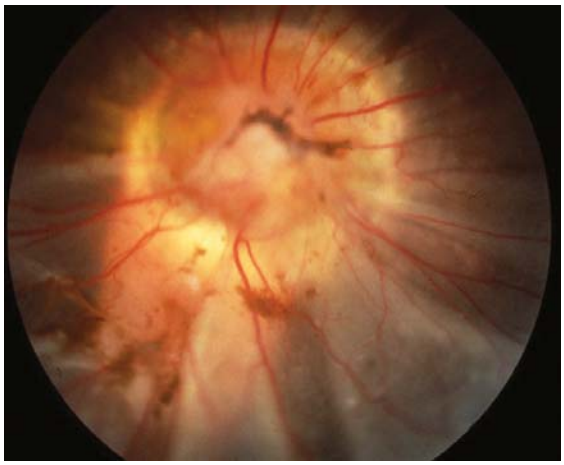

Fig. 19.4: Morning glory syndrome
(*Courtesy*: Mr S Kanagami, Tokyo)

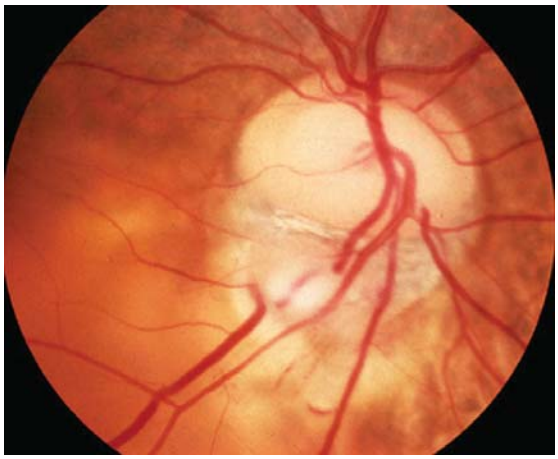

Fig. 19.3: Congenital pit of optic disk

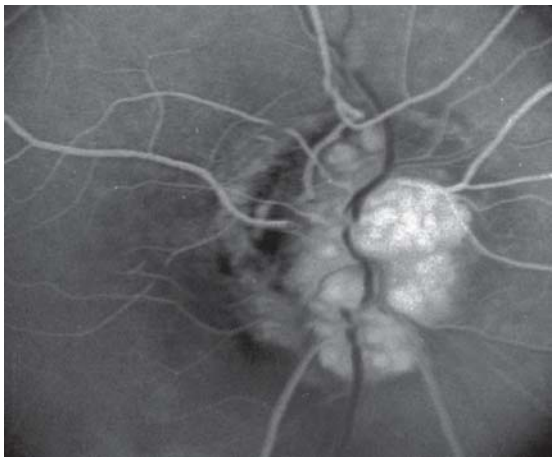

Fig. 19.5: FA of drusen of optic disk
(*Courtesy*: Mr S Kanagami, Tokyo)

resembling the morning glory flower (Fig. 19.4). The characteristic features include an enlarged optic disk containing persistent hyaloid remnants, peripapillary pigmentary changes, emergence of retinal vessels from the periphery of the disk and nonrhegmatogenous retinal detachment. The vision is usually defective.

Optic Disk Drusen

Drusen of the optic disk (Fig. 19.5) is usually bilateral and familial. It is characterized by the absence of the optic cup, presence of spontaneous venous pulsations and abnormal branching of the retinal vessels from the center of the disk. It may be confused with early papilledema.

VASCULAR DISTURBANCES OF THE OPTIC NERVE

Papilledema or Edema of the Optic Nerve Head (Choked Disk)

A noninflammatory swelling of the optic nerve head is known as *papilledema*.

Theories of Papilledema

The pathogenesis of papilledema is disputed. Two main etiological theories have been advanced:

1. Compression of the central retinal vein, and
2. Stasis of axoplasm.

The optic nerve is enclosed within the meningeal sheaths common to the brain. The raised intracranial pressure produces distension of the intravaginal space around the nerve and causes compression of the central retinal vein while it crosses the subarachonoid space. Earlier it was accepted as the most probable mechanism of development of papilledema. However, it lacked experimental evidence.

Hayreh experimentally demonstrated that papilledema develops due to blockage of the axonal transport. The raised intracranial pressure causes interruption of the axoplasmic flow at the level of lamina cribrosa leading to swelling of the optic disk and vascular changes at and around the optic nerve head.

Pathology

Papilledema presents a noninflammatory swelling of the optic nerve head accompanied with peripapillary edema of the nerve fiber layer, and dilatation of disk surface capillary net and retinal veins associated with peripapillary hemorrhages and exudates. The edema often throws the internal limiting membrane into folds and obliterates the physiological cup. The nerve fiber layer degenerates and multiple colloid bodies appear on the lamina cribrosa. In late phase of papilledema, proliferation of neuroglia occurs and the mesoblastic tissue around the blood vessels get thickened.

Etiology

Papilledema may result from a number of conditions including intracranial space occupying lesions, hydrocephalus, meningitis, cerebral venous obstruction and intracranial hemorrhage.

Table 19.1: Causes of unilateral and bilateral papilledema

Unilateral papilledema	*Bilateral papilledema*
1. *Ocular conditions* Central retinal vein occlusion Ischemic optic neuropathy Ocular hypotonia	1. *Intracranial tumors* Midbrain tumors Parieto-occipital tumors Cerebellar tumors
2. *Orbital lesions* Orbital cellulitis Orbital venous thrombosis Orbital tumors Meningioma of optic nerve Carotico-cavernous fistula Metastatic orbital masses Early thyroid ophthalmopathy Hemorrhage in optic nerve sheath Pseudotumors	2. *Other intracranial lesions* Aneurysms Thrombosis of cavernous sinus (late) 3. Systemic diseases Malignant hypertension Nephritis Toxemia of pregnancy Blood dyscrasias Giant cell arteritis Late thyroid ophthalmopathy
3. *Intracranial lesions* Posterior fossa tumors Brain abscess Early cavernous sinus thrombosis Pseudotumor cerebri Foster-Kennedy syndrome Tumor of orbital surface of frontal lobe Olfactory groove meningioma	

Systemic diseases like malignant hypertension, nephritis, toxemia of pregnancy and blood dyscrasias may be associated with bilateral papilledema. Unilateral papilledema occurs in orbital lesions such as orbital tumors or abscess. The causes of unilateral and bilateral papilledema are listed in Table 19.1.

Clinical Features

Visual symptoms of papilledema usually occur late since the vision remains unimpaired for a long time. Transient attacks of blurred vision or blackouts lasting for a few minutes to an hour

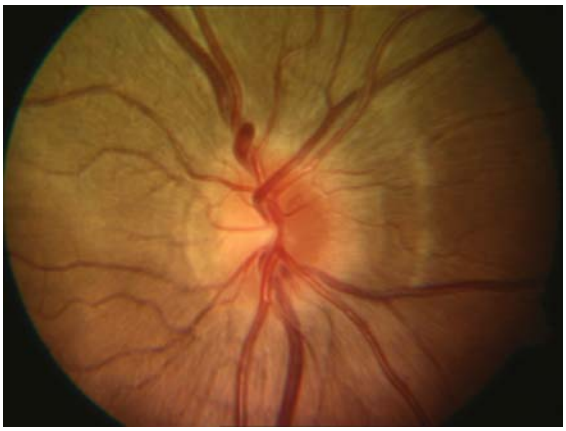

Fig. 19.6: Early papilledema (*Courtesy*: Dr T Sharma, Sankara Nethralaya, Chennai)

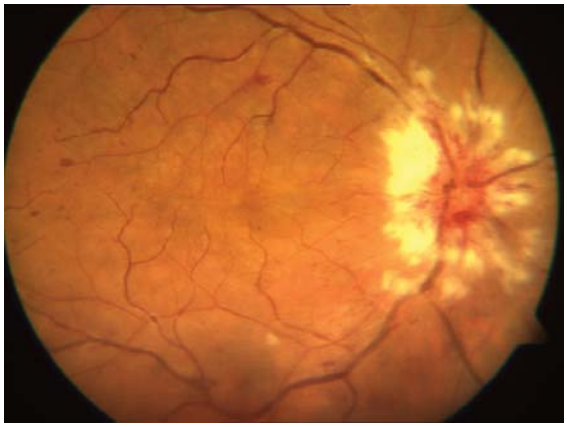

Fig. 19.7: Acute phase of papilledema (*Courtesy*: Dr T Sharma, Sankara Nethralaya, Chennai)

may occur. The vision is affected either by macular edema/exudates or with the onset of optic atrophy. Constitutional symptoms like persistent headache, nausea and vomiting are frequent.

Papilledema may present following four phases:

Early Phase: Hyperemia of the disk, congestion of veins, absence of venous pulsations and slight blurring of the disk margins (Fig. 19.6) are the early signs of papilledema. The blurring starts at the nasal margin initially, and then upper, lower and temporal margins get affected and become indistinct.

Initially, the physiological cup is preserved (a feature which distinguishes papilledema from the optic neve drusen). The edema spreads and produces concentric or radial peripapillary retinal folds known as *Paton lines*. The retinal veins become tortuous and markedly dilated. The vascular engorgement and stasis lead to extensive flame-shaped and punctate hemorrhages, particularly marked around the disk.

Acute Phase: Visual acuity, color vision and pupillary reactions are often normal. However, the patient may complain of transient attacks of blackouts of vision associated with headache, nausea and vomiting. The optic cup is usually filled and peripapillary flame-shaped hemorrhages and cotton-wool spots are marked (Fig. 19.7). The edema may spread and produce folds around the macula with multiple scattered exudates in the retina, sometimes arranged in a radial manner forming an incomplete macular star or fan (Fig. 19.8).

Chronic Phase: The disk may not be hyperemic but appear pale (Fig. 19.9). Small refractile bodies may be present on the surface of the disk. The optic cup remains obliterated and the optic nerve head resembles the dome of a champagne cork. The

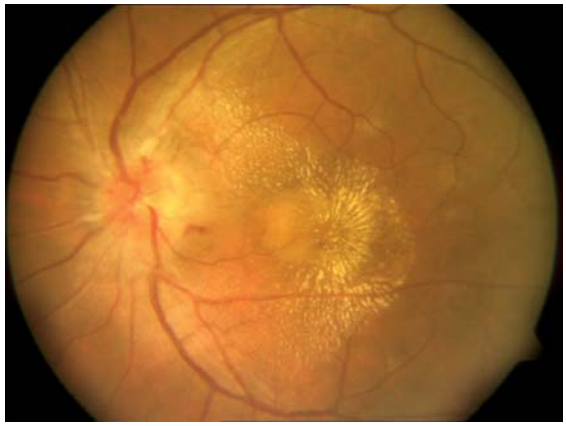

Fig. 19.8: Acute phase of papilledema with macular star (*Courtesy*: Dr T Sharma, Sankara Nethralaya, Chennai)

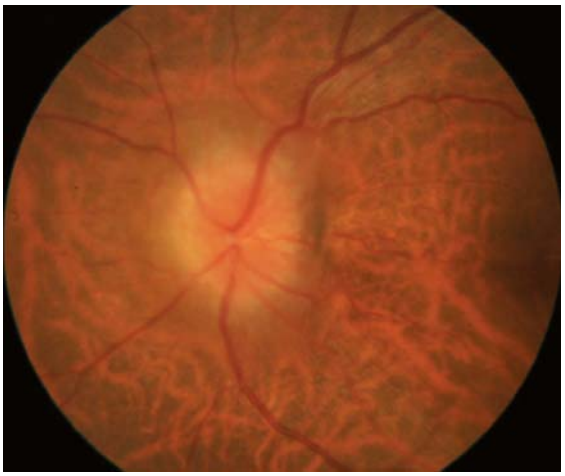

Fig. 19.9: Chronic phase of papilloedma (*Courtesy*: Mr S Kanagami, Tokyo)

peripapillary nerve fiber layer appears grayish due to gliosis and the retinal vessels show sheathing.

Atrophic Phase: It is characterized by reactive proliferation of astrocytes resulting in post-papilledematous optic atrophy.

Differential Diagnosis

Pseudopapillitis mimics papilledema. Pseudopapillitis is seen in high hypermetropic eye in which the porus opticus is small and the optic nerve fibers are heaped up causing blurring of the disk margin. Venous engorgement, hemorrhages and exudates are not present. Papilledema must be differentiated from pseudopapilledema and drusen of the optic disk. The distinguishing features are listed in Table 19.2.

The papilledema should also be differentiated from papillitis, an inflammatory condition.

Management

The management of papilledema is essentially the treatment of the cause. Prompt control of raised intracranial pressure resolves papilledema and may not leave any permanent visual defect. But if the condition persists, postpapilledematous optic atrophy develops and permanent blindness ensues.

Table 19.2: Differentiating features of papilledema, pseudopapilledema and optic disk drusen

Features	*Papill-edema*	*Pseudopapill-edema*	*Optic drusen*
Size of optic disk	Normal or enlarged	Small	Small
Optic cup	Initially preserved	Absent	Absent
Color of optic disk	Hyperemic	Normal	Pale
Venous pulsation	Absent	May be present	May be present
Peripapillary nerve fibers	Edematous	Normal	Normal
Peripapillary hemorrhages	Present	Absent	Absent
Retinal veins	Dilated and tortuous	Normal	Normal
FA	Early leakage of dye	Normal	Intrinsic fluorescence

Anterior Ischemic Optic Neuropathy

Anterior ischemic optic neuropathy (AION) is an acute optic neuropathy in patients of about the age of 50 years. It is classified into two types.

1. Arteritic anterior ischemic optic neuropathy (AAION), and
2. Nonarteritic anterior ischemic optic neuropathy (NAION).

Arteritic Anterior Ischemic Optic Neuropathy

Etiology

Arteritic anterior ischemic optic neuropathy is less frequent and predominantly affects old females (mean age 70 years). It is caused by inflammatory

and thrombotic occlusion of the short posterior ciliary arteries. It is often associated with giant cell arteritis.

Clinical Features

Headache and tenderness of temporal artery on skull are common features. Visual loss is severe. Signs of retinal ischemia include pale blurred disk, cotton-wool spots and retinal edema.

Diagnosis: Raised ESR (70-100 mm/hour), abnormal C-reactive protein and temporal artery biopsy often confirm the diagnosis. Visual field defects are extensive and include altitudinal or arcuate scotomas.

Treatment

Treatment must be started immediately to prevent contralateral visual loss. Intravenous methyl prednisolone is recommended 1 g/day for 3-5 days followed by 100 mg/day oral prednisolone; it should be tapered over 3-12 months.

Nonarteritic Anterior Ischemic Optic Neuropathy

Etiology

Nonarteritic anterior ischemic optic neuropathy occurs in relatively younger age group (mean age 60 years) and is caused by compromise in the optic disk microcirculation (crowding of the disk). Hypertension, diabetes, smoking, systemic lupus erythematosus and migraine are known risk factors.

Clinical Features

Visual impairment on awakening is a common symptom. Optic disk edema (Fig. 19.10) is diffuse or segmental and is associated with telangiectasia of the disk, flame-shaped hemorrhages, hard exudates and cotton-wool spots. Altitudinal visual field loss occurs mostly in the inferonasal quadrant (Fig. 19.11). The presence of the small optic disk without a physiological cup (disk at risk) in the contralateral eye is a bad sign.

Treatment

There is no effective treatment for NAION. Hyperbaric oxygen, optic nerve sheath decompression, systemic aspirin and levodopa have been tried with variable success.

INFLAMMATION OF THE OPTIC NERVE (OPTIC NEURITIS)

Optic neuritis is a broad term encompassing inflammatory and demyelinating disorders of any part of the optic nerve.

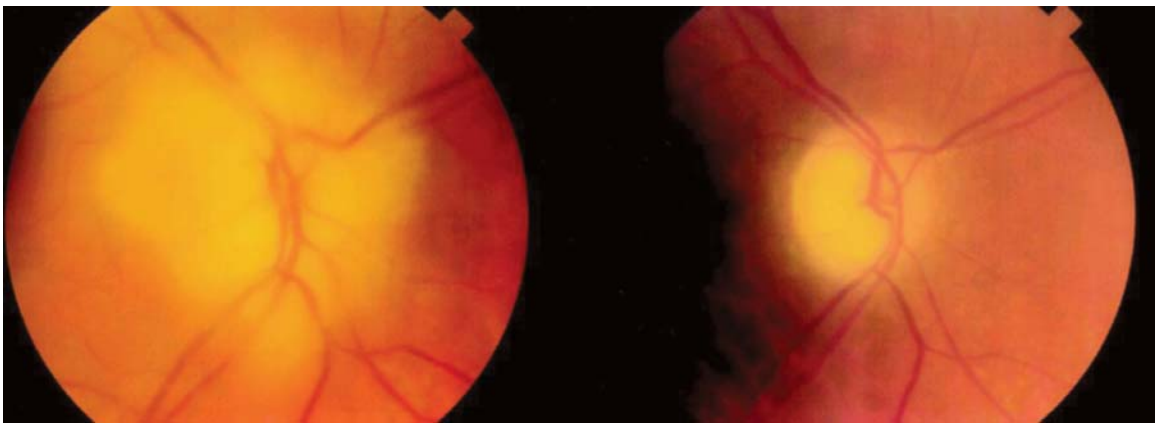

Fig. 19.10: Anterior ischemic optic neuropathy (*Courtesy*: Dr. G Chandra Sekhar, LVPEI, Hyderabad)

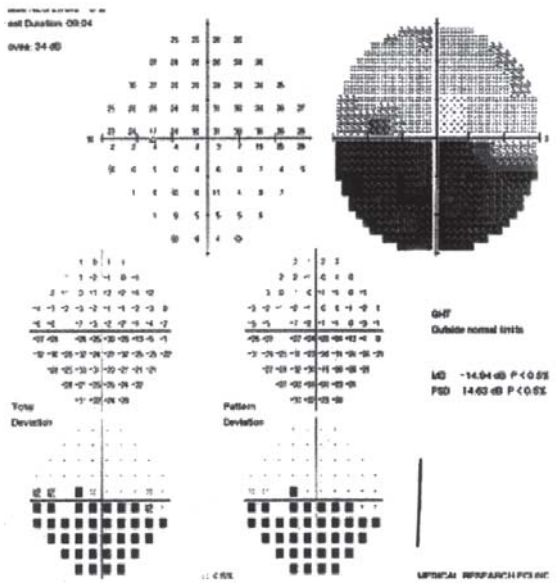

Fig. 19.11: Visual field defect in AION
(*Courtesy*: Sankara Nethralaya, Chennai)

Clinically, optic neuritis is divided into three groups:

1. *Papillitis* (inflammation of the intraocular part of the nerve)
2. *Retrobulbar neuritis* (inflammation of the retrobulbar part of the nerve), and
3. *Neuroretinitis* (inflammation of the optic nerve and the retina).

Etiology

Optic neuritis occurs in a number of systemic and ocular diseases:

1. Demyelinating disease is the most common cause, particularly multiple sclerosis
2. Viral infection: poliomyelitis, herpes, chicken pox, mumps and measles
3. Systemic granulomatous inflammation such as tuberculosis, sarcoidosis neurosyphilis and neuromyelitis
4. Secondary involvement of optic nerve in meningitis, sinusitis, orbital cellulitis and retinochoroiditis
5. Autoimmune vascular disorders such as systemic lupus erythematosus and polyarteritis nodosa can induce optic neuritis secondary to ischemia
6. Lebers hereditary optic neuropathy
7. Toxic
8. Idiopathic.

Nutritional and metabolic disorders such as pernicious anemia, diabetes mellitus and hyperthyroidism may be considered as risk factors.

Papillitis

Papillitis is an inflammation of the intraocular part of the optic nerve.

Clinical Features

Papillitis is often unilateral and loss of vision is the hallmark of the disease. There may be pain on ocular movements and the pupillary light reflex is sluggish. The patient may complain of a depressed light-brightness and fading of colored objects.

Papillitis usually presents an indistinguishable ophthalmoscopic picture from papilledema. The disk is hyperemic and swollen with blurred margins (Fig. 19.12). Extensive peripapillary retinal edema may be present. The veins are

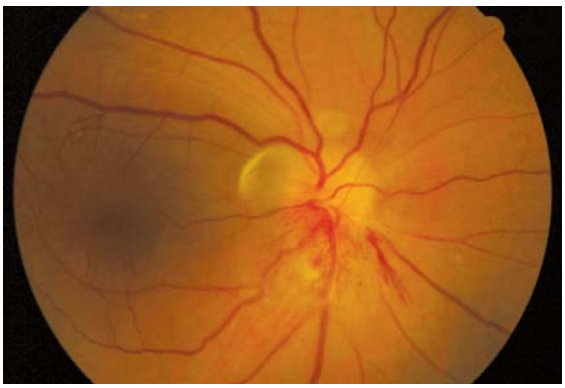

Fig. 19.12: Papillitis

dilated and tortuous, hemorrhages and exudates appear upon the disk and in the retina, and there are fine vitreous cells opacities. Central or centrocecal scotoma is a common visual field defect.

Cases of mild papillitis may recover completely but severe affection often leads to postneuritic optic atrophy. The latter is characterized by a dirty gray colored disk with filled cup and indistinct margin owing to glial proliferation, and perivascular sheathing of the vessels.

Papillitis should be differentiated from papilledema. The important distinguishing clinical features are listed in Table 19.3.

Table 19.3: Differentiation between papillitis and papilledema

Features	*Papillitis*	*Papilledema*
Laterality	Usually unilateral	Usually bilateral
Onset	Generally sudden	Generally insidious
Loss of vision	Sudden and marked	Gradual and negligible in the initial stage
Pain	Present on ocular movements	Absent
Tenderness	Present at the insertion of superior rectus	Absent
Swelling of the disk	Moderate (2-3 diopters)	Marked (more than 3 diopters)
Visual field defects	Central or centrocecal scotoma	Enlargement of blind spot and concentric constriction
Posterior vitreous	Fine cells opacities	Clear

Retrobulbar Neuritis

Retrobulbar neuritis is an inflammation of the retrobulbar part of the optic nerve.

On the basis of onset, the retrobulbar neuritis is usually divided into an acute and a chronic form.

The acute form manifests when there is primary involvement of the nerve fibers, while in chronic form, degeneration of nerve fibers occurs probably due to damage to the ganglion cells of the retina from the absorption of exogenous toxins. Therefore, it is also known as *toxic amblyopia*.

Acute Retrobulbar Neuritis

Clinical Features

The disease is usually unilateral and starts with sudden and marked loss of vision. It may be associated with headache and neuralgia. Ocular movements are painful, especially in upward and inward directions. The pain increases by pressure upon the globe. There is a local tenderness in the region of the insertion of superior rectus muscle.

The pupil reacts to light but the constriction is not sustained, it slowly dilates even in the presence of bright light. Such a reaction is known as *ill-sustained pupillary reaction* (*Marcus-Gunn pupil*). Besides the pupillary abnormality, other visual functions are also altered. The colored object may look washed-out and there is a depression of light-brightness. The depth perception particularly for moving objects is impaired (*Pulfrich's phenomenon*).

Ophthalmoscopy does not reveal any obvious sign unless the lesion is close to the lamina cribrosa. Occasionally temporal pallor of the ONH may be seen. Central or centrocecal scotoma is often found due to the involvement of papillomacular bundle.

Course

The course of the disease is rapidly progressive and in some cases complete blindness may develop within weeks. In majority of the cases, there is a spontaneous and more or less complete recovery. But the disease has a tendency for remissions. It may cause a partial optic atrophy which is characterized by the presence of temporal pallor of the disk and central or centrocecal scotoma.

Differential Diagnosis

Retrobulbar neuritis should be differentiated from hysterical and cortical blindness. Magnetic

resonance imaging (MRI) studies of brain and orbits should be ordered to look for demyelination of the central nervous system.

Treatment

The treatment of optic neuritis is basically the treatment of its cause. Systemic administration of corticosteroids helps in the speedy recovery of vision. Oral corticosteroid therapy alone may increase the risk of recurrence of optic neuritis.

The patients with optic neuritis need observation as recovery in the natural course is the rule. In the first episode of optic neuritis with no history of multiple sclerosis and MRI confirmation of demyelination, the Optic Neuritis Treatment Trial (ONTT) recommends the use of pulsed methyl prednisolone 1 g intravenous daily for 3 days followed by oral prednisolone 1 mg/kg body weight daily for 11 days. The oral prednisolone is quickly tapered and stopped in next 3 days. In the patients of multiple sclerosis with optic neuritis or in recurrent attacks of optic neuritis, observation is advised.

Neuroretinitis

Papillitis associated with retinitis is called *neuroretinitis.* It occurs in children and is more often bilateral.

Neuroretinits often develops following viral infection, cat-scratch fever and Lyme disease. It is not a manifestation of demyelinating disease. Neuroretinitis presents with features of papillitis associated with a characteristic macular star with multiple exudates. The treatment of neuroretinitis is same as that of optic neuritis. However, in most cases it is a self-limiting disease which resolves in 6-12 months after appropriate treatment of the primary disease.

Leber's Hereditary Optic Neuropathy

Leber's hereditary optic neuropathy (LHON) is a form of retrobulbar neuritis manifesting at about 20 year of age predominantly affecting males.

Etiology

LHON is related to a mitochondrial DNA mutation most commonly at 11778 position. The condition is transmitted in a sex-linked manner generally through unaffected females to males.

Clinical Features

The disease is characterized by unilateral or bilateral rapid deterioration of vision which later becomes stationary or even shows improvement. The patient often has defective color perception.

Diagnosis

The diagnosis of LHON is based on its familial and hereditary character, pallor of the disk and the presence of centrocecal scotoma. In some cases the disk margins may be found blurred.

Pseudoedema of the disk, peripapillary telangiectasis and non-leakage of fluorescein from the telangiectatic vessels on fluorescein angiography (FA) constitute the classical triad of Leber's optic neuropathy.

Most of the cases on visual field recording develop a central or centrocecal scotoma and a few present with concentric contraction of the field or peripheral sector-shaped defects.

Treatment

Earlier visual improvement had been claimed after the use of cyanocobalamine. However, no treatment has been shown to be effective.

Toxic Amblyopia

Toxic amblyopia or chronic retrobulbar neuritis includes a number of entities in which the optic nerve fibers are damaged by exogenous toxins. Certain toxins have a direct effect on the nerve fibers while others, such as tobacco and methyl alcohol, primarily affect the ganglion cells of the retina and cause secondary degeneration of the nerve fibers. The common poisons which lead to toxic amblyopia are tobacco, ethyl alcohol, methyl

alcohol, lead, arsenic, quinine, carbon disulphide and Cannabis indica. Some of the antitubercular drugs (ethambutol, isonicotinic hydrazide), antimalarials (chloroquine, quinine) and a few other chemotherapeutic agents (amiodarone, vigabatrin, sulfonamides, digitalis, chlorpropamide, tolbutamide) are neurotoxic and may induce toxic amblyopia.

Tobacco Amblyopia

Etiology

An abuse of tobacco either by smoking or chewing can cause toxic amblyopia. Its toxicity increases if the patient also has an over indulgence in alcohol or suffers from nutritional deficiency, particularly of vitamin B-complex. The tobacco contains nicotine and its volatile decomposed products, collidine and lutidine, which are toxic. Cyanide present in tobacco smoke is extremely toxic and injurious.

Clinical Features

The patient usually complains of diminution of vision and difficulty in near work. A history of consumption of tobacco, temporal pallor of the disk, color confusion and presence of centrocecal scotoma are helpful in the diagnosis of tobacco amblyopia.

Treatment

Complete abstinence from tobacco, large doses of vitamin B_1, B_6 and B_{12} and systemic vasodilators may improve the vision.

Methyl Alcohol Amblyopia

Etiology

Methyl alcohol amblyopia occurs from drinking methylated spirit which is oxidized into formic acid and formaldehyde causing swelling and degeneration of the ganglion cells of the retina.

Clinical Features

The fundus examination may reveal optic atrophy. Nausea, vomiting, giddiness and coma are constitutional features.

Treatment

Gastric lavage and administration of intravenuos sodabicarbonate, folic acid and ethyl alcohol may help the patient.

Quinine Amblyopia

Etiology

Quinine amblyopia may occur following the use of quinine even in small doses in susceptible persons. The recommended dose of the drug is 150 mg per day.

Clinical Features

The patient develops a near total blindness associated with tinnitus and deafness. The pupils are dilated and fixed. Marked pallor of the optic disk, extreme attenuation of retinal vessels and retinal edema are characteristic ophthalmoscopic signs.Visual fields show marked constriction. Permanent blindness may develop due to optic atrophy.

Treatment

Elimination of the drug, administration of multivitamins (B_1, B_6, B_{12}) and nutritional supplementation may restore some useful vision.

Ethambutol Amblyopia

Etiology

Ethambutol is commonly used in the treatment of tuberculosis. It is administered in the doses of 15 mg/kg/day. Toxicity of ethambutol is likely to occur in alcoholic and diabetic patients.

Clinical Features

Some patients may notice defective color vision and reduced visual acuity during the course of treatment. The drug may induce edema of the optic nerve head, splinter hemorrhages and optic neuritis. Central or centrocecal scotoma is a typical visual field defect but when the optic chiasma is involved, bitemporal hemianopia develops.

Treatment

Withdrawal of the drug and administration of vitamin B-complex, vitamin C and zinc may improve the vision.

DEGENERATION OF THE OPTIC NERVE (OPTIC ATROPHY)

Injury to the optic nerve fibers in any part of the course from the retina to the lateral geniculate nucleus leads to their degeneration and subsequent optic atrophy.

Classification of Optic Atrophy

Optic atrophy can be classified on the basis of ophthalmoscopic appearance as primary and secondary.

Primary Optic Atrophy

In primary optic atrophy the disk is white with a bluish-tint, lamina cribrosa is seen, margin is sharply defined and retinal blood vessels and surrounding retina appear normal (Fig. 19.13). This type of optic atrophy reflects a chronic process and is not usually preceded by congestion or edema of the optic disk. Hydrocephalus, intracranial meningioma and tabes dorsalis may produce a primary optic atrophy.

Secondary Optic Atrophy

In secondary optic atrophy the optic disk is waxy gray, lamina cribrosa is not seen and cup is filled,

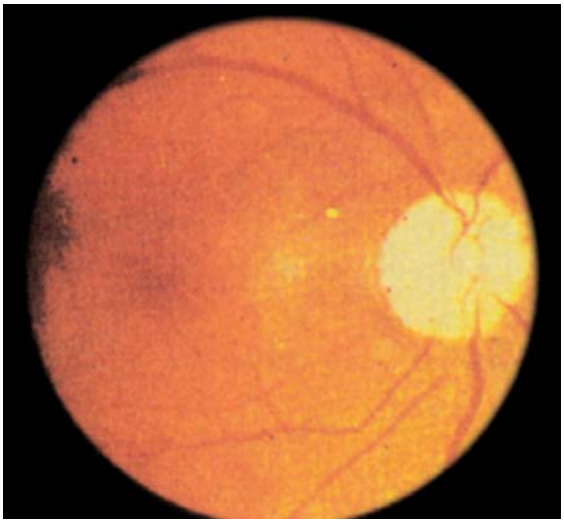

Fig. 19.13: Primary optic atrophy (*Courtesy*: Dr SS Badrinath, Chennai)

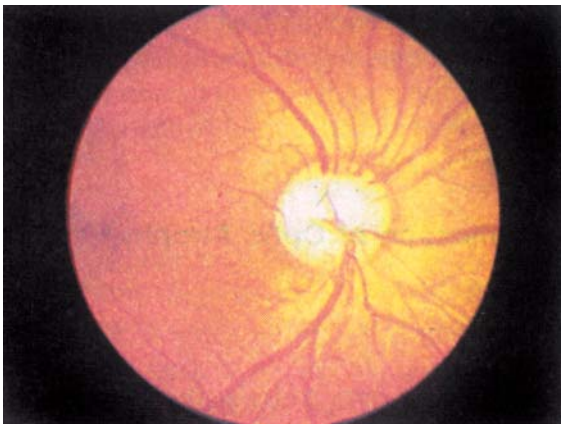

Fig. 19.14: Optic atrophy secondary to retinitis pigmentosa (*Courtesy*: Mr S Kanagami, Tokyo)

the edges are blurred, retinal vessels are attenuated and the retina is unhealthy (Fig. 19.14).

Optic atrophy can also be classified as ascending and descending types.

Ascending Optic Atrophy

In ascending type of optic atrophy, the primary lesion is in the retina or the optic disk. It occurs

due to glaucoma, retinochoroiditis, retinitis pigmentosa and central retinal artery occlusion. They cause a secondary effect on the optic nerve and white tracts in the brain.

Descending Optic Atrophy

In descending type of optic atrophy, the primary lesion usually lies in the brain or in the optic nerve. Intracranial space occupying lesions, meningitis and demyelinating diseases are common causes of descending optic atrophy.

The above classifications of optic atrophy are confusing and not clinically helpful. Therefore, a classification of optic atrophy based on the etiology is preferred.

Etiological Classification of Optic Atrophy

1. *Congenital hereditary optic atrophy* may appear at birth or manifest later in life (Behr's optic atrophy).
2. *Consecutive optic atrophy* occurs secondary to the retinal diseases such as retinitis pigmentosa (Fig. 19.14), high myopia, retinal detachment and retinochoroiditis.
3. *Vascular optic atrophy* occurs due to occlusion of central retinal artery or vein, arteriosclerosis, anemia and sudden massive loss of blood.
4. *Postpapilledematous optic atrophy* is a sequel to long-standing papilledema.
5. *Postneuritic optic atrophy* ensues after optic neuritis (Fig. 19.15).
6. *Pressure optic atrophy* occurs due to pressure on optic chiasma by pituitary tumors, aneurysm of the circle of Willis or focal basal arachnoiditis. Nerve can also be strangulated at the optic foramen in osteitis deformans.

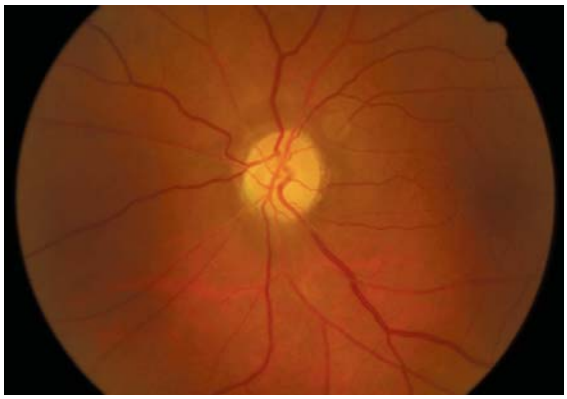

Fig. 19.15: Postneuritic optic atrophy

7. *Traumatic optic atrophy* results from a direct injury to the nerve or its nutrient vessels.
8. *Toxic optic atrophy* is a common feature of toxic amblyopia.
9. *Metabolic optic atrophy* may be found in diabetes mellitus and gangliosidosis.
10. *Glaucomatous optic atrophy* develops in long-standing cases of glaucoma and is characterized by marked excavation of the optic cup.

Pathology

Damage to the retinal ganglion cells is the hallmark of optic atrophy. Histologically, there occurs a loss of axons and supporting connective tissues of the optic nerve with variable degrees of glial proliferation. In primary optic atrophy, there is a relative absence of mesenchymal and glial tissues, but in secondary optic atrophy gliosis and proliferation of astrocytes on and about the optic disk impart it a dirty gray appearance.

Clinical Features

The optic nerve functions are usually deranged in optic atrophy. The nerve functions can be assessed

by visual acuity and color vision, pupillary reactions, optic disk appearance and visual fields.

Vision: Mild or severe loss of vision is the main symptom of optic atrophy. The impairment of vision may be sudden or gradual depending on the etiology. Color perception is also impaired.

Pupils: Both direct and consensual pupillary reactions are absent in bilateral optic atrophy. However, in unilateral optic atrophy, there is a loss of ipsilateral direct and contralateral consensual pupillary reactions.

Optic disk: Mild form of optic atrophy may present with normal orange-pink disk color or subtle pallor with thin or absent surface vascular net of the disk. The number of small blood vessels on the optic nerve head may be reduced (*Kestenbaum sign*). Severe form of optic atrophy is identified by a chalky-white appearance of the optic disk with well-defined margins. Disk looks waxy in consecutive optic atrophy. The optic disk appears dirty pale with blurred margins and filled cup in postneuritic optic atrophy and postpapilledematous optic atrophy.

Nerve fiber layer: Slit or rake defects, sector atrophy and diffuse atrophy in the peripapillary nerve fiber layer and papillomacular bundles may precede optic atrophy.

Visual fields: Visual field defects are important features of optic atrophy. Central or peripheral scotomatous defects may be found depending on the area of damage of optic nerve fibers.

Treatment

The treatment of optic atrophy is largely preventive. The underlying cause of optic atrophy must be treated in early stages to prevent the nerve damage. Prognosis is usually poor once optic nerve is completely damaged.

TUMORS OF THE OPTIC NERVE

The tumors of the optic nerve are classified as primary and secondary.

The primary tumors of the optic nerve include:

1. Glioma or astrocytoma
2. Oligodendrocytoma
3. Meningioma
4. Melanocytoma
5. Hemangioma
6. Medulloepithelioma.

Glioma, meningioma, melanocytoma and hemangioma are described in the *chapter on Diseases of the Orbit*.

The optic nerve is secondarily involved in retinoblastoma, malignant melanoma of the choroid and intracranial meningioma. Lymphoma and leukemia can also involve the optic nerve.

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CHAPTER

20 Lesions of the Visual Pathway

The visual pathway consists of retina, optic nerve, optic chiasma, optic tract, lateral geniculate nucleus, optic radiations and visual cortex. The localization of lesions of the visual pathway has a great importance in neuro-ophthalmology (Fig. 20.1).

LESIONS OF THE VISUAL PATHWAY

Lesions of the Retina

Visual space is represented on the retina in direct point-to-point relationship. The imaginary line dividing nasal and temporal fibers passes through the center of fovea. All temporal fibers lying lateral to the line do not cross, while the nasal fibers cross to the opposite optic tract in the chiasma. The fibers of the upper and lower halves of the peripheral retina do not overlap. Because of this orderly arrangement, the superior visual field is projected onto inferior retina, the nasal field onto temporal retina, the inferior visual field onto superior retina and the temporal field onto nasal retina. Owing to this inverted relationship, lesions of the retina cause defects in the corresponding opposite visual field. When papillomacular fibers are involved the central vision is affected.

Lesions of the Optic Nerve

Involvement of both the optic nerves causes complete blindness with the absence of pupillary

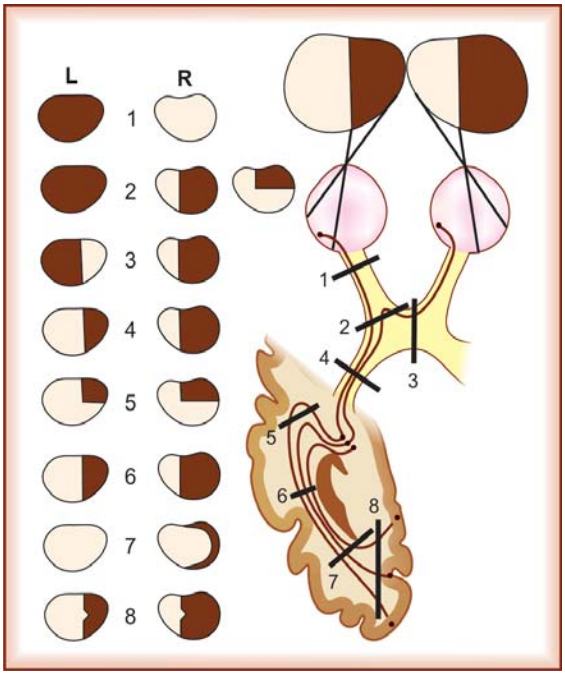

Fig. 20.1: Lesions of the visual pathway and corresponding field defects:

1. Lesion through optic nerve—ipsilateral blindness
2. Lesion through proximal part of optic nerve—ipsilateral blindness with contralateral hemianopia
3. Sagittal lesion of chiasma—bitemporal hemianopia
4. Lesion of optic tract—homonymous hemianopia
5. Lesion of temporal lobe—quadrantic homonymous defect
6. Lesion of optic radiations—homonymous hemianopia (sometimes sparing the macula)
7. Lesion in anterior part of occipital cortex—contralateral temporal crescentic field defect
8. Lesion of occipital lobe—homonymous hemianopia (usually sparing the macula)

reaction to light. If only one optic nerve is damaged, it results in ipsilateral blindness with loss of ipsilateral direct and contralateral consensual pupillary reactions.

The lesion of the proximal part of optic nerve results in ipsilateral blindness and contralateral superotemporal field defect (*Traquair junctional scotoma*) due to looping of crossed fibers in the optic nerve of opposite side.

Lesions of the Optic Chiasma

The nasal fibers, which constitute about 60% of the total fibers, cross in the chiasma to the opposite optic tract. The temporal fibers do not cross and run in the optic tract of the same side. The fibers from the lower and nasal quadrants of the retina bend medially into the anterior portion of chiasma. After crossing, the anterior fibers in the chiasma loop into the optic nerve of opposite side.

The chiasmal fibers, because of their closeness to pituitary gland, are liable to be compressed by enlargement of the gland resulting in optic atrophy (Fig. 20.2). The chiasmal tumors cause characteristic visual field defects in about 80% cases. As one side is usually compressed before the other, the earliest defect is a unilateral scotoma. It may be followed by homonymous hemianopia (Fig. 20.3) due to pressure on one optic tract or occasionally an altitudinal hemianopia (loss of upper or rarely the lower half of the field). An intra-sellar or extra-sellar tumor produces pressure upon the chiasma and causes early loss in the upper half of field, while supra-sellar tumors cause early loss in the lower half of visual field, and later bitemporal hemianopia may develop.

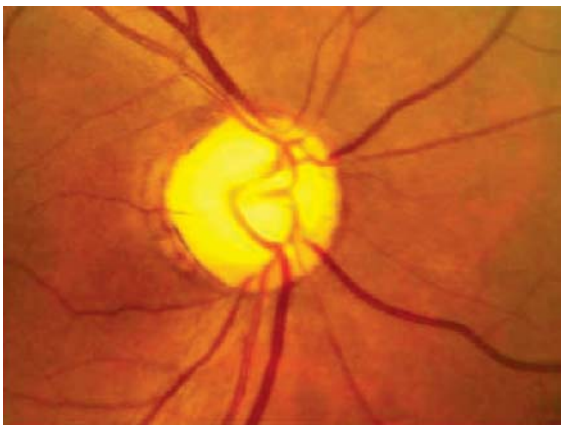

Fig. 20.2: Optic atrophy caused by pituitary adenoma

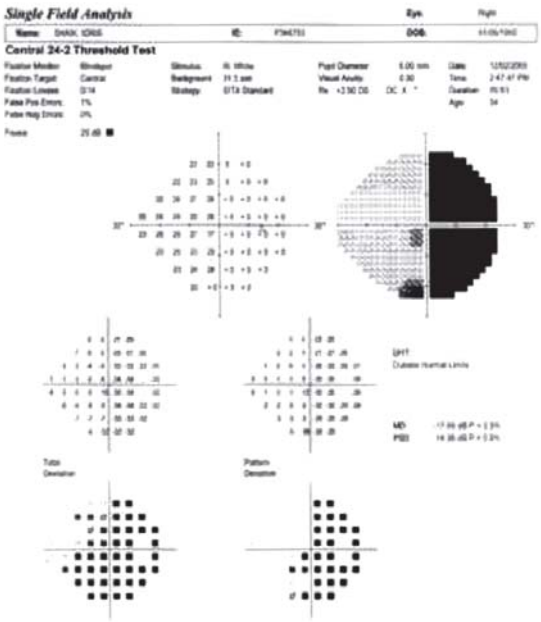

Fig. 20.3: Pituitary adenoma causing temporal hemianopic visual field defect

Loss of one half of field of vision is known as *hemianopia*. When there is loss of the temporal half of one field of binocular vision and the nasal half of the other field the condition is called as *homonymous hemianopia*.

Bitemporal hemianopia (Fig. 20.4) is usually produced by tumors of sella turcica, suprasellar aneurysms and chronic arachnoiditis by pressing upon the chiasma and destroying the fibers of nasal halves of each retina.

Binasal hemianopia (Fig. 20.5), although rare, is caused by the enlargement of third ventricle and atheromas of the carotids or posterior communicating arteries, because they destroy the fibers of temporal halves of each retina.

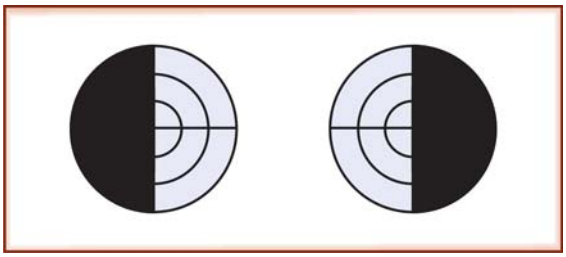

Fig. 20.4: Bitemporal hemianopia

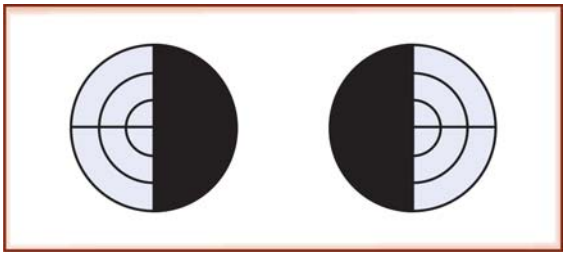

Fig. 20.5: Binasal hemianopia

Lesions of the Optic Tract

The optic tract carries uncrossed temporal fibers of the same side and crossed nasal fibers of the opposite side, therefore, a lesion of the tract results in homonymous hemianopia. As the arrangement of the nerve fibers in the tract is not regular, lesions of the tract give *incongruous* (two sides not exactly equal) *homonymous hemianopia*.

The afferent pupillary fibers (20%) accompany the visual fibers in the optic tract and reach the pretectal area in the midbrain where they synapse. When light falls on the blind halves of the retina in patients with homonymous hemianopia, the pupils do not react, but when it falls on the other halves of the retina they react, a condition called *Wernicke's hemianopic pupillary reaction*.

The association of hemianopia with contralateral third cranial nerve palsy and ipsilateral hemiplegia indicates an optic tract lesion.

Syphilitic and tuberculous meningitis, tumors of optic thalamus, tentorial meningioma and aneurysm of the superior cerebellar and posterior cerebral arteries cause optic tract lesions. They cause involvement of fixation point and bilateral partial optic atrophy.

Lesions of the Lateral Geniculate Body

The lateral geniculate bodies (LGB) are ovoid and situated at the posterior end of the optic tract. The fibers of the optic tract enter the LGB and the optic radiations originate from there. Geniculate or infrageniculate lesions cause homonymous hemianopia.

Lesions of the Optic Radiations

The visual fibers running between the lateral geniculate bodies and the occipital lobe constitute the optic radiations (Fig. 20.6). They originate

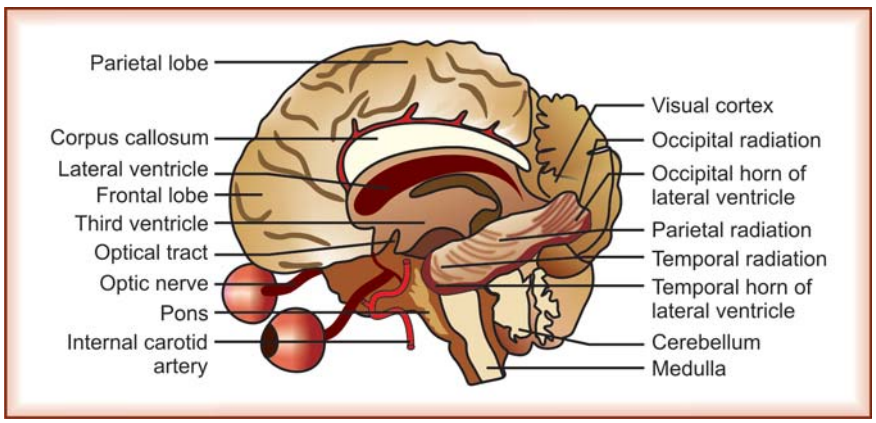

Fig. 20.6: Diagram showing optic radiation

from the lateral geniculate body, pass through the posterior part of the internal capsule and terminate around the calcarine fissure in the occipital lobe of brain. The latter is also known as *visual cortex*.

The arrangement of fibers in the optic radiation is systematic. The fibers from the temporal upper quadrant of the ipsilateral and the nasal upper quadrant of the contralateral retina are present in the upper half of the radiation, while the lower half of the radiation represents the lower quadrants of the corresponding retina. As the nerve fibers in the optic radiations are regularly arranged, the lesions of the optic radiations (brain abscess, tumors and vascular lesions) give *congruous homonymous hemianopia*.

The most inferior fibers of optic radiation pass anterolaterally and then posteriorly to loop around the temporal horn of the lateral ventricles (Meyer's loop). The loop lies approximately 2.5 cm from the anterior tip of the temporal lobe. More superiorly, the visual fibers travel posteriorly through the parietal lobe of brain.

The lesions affecting Meyer's loop in the temporal lobe produce superior homonymous quadrantic defects ("pie in the sky") on the opposite side sparing the fixation area (Fig. 20.7). The lesion of the temporal lobe causes complete superior homonymous quadrantanopia (Fig. 20.8).

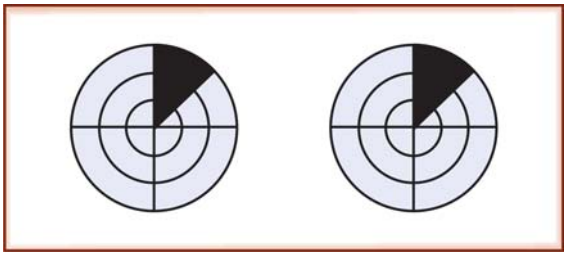

Fig. 20.7: Right superior homonymous quadrantic defect

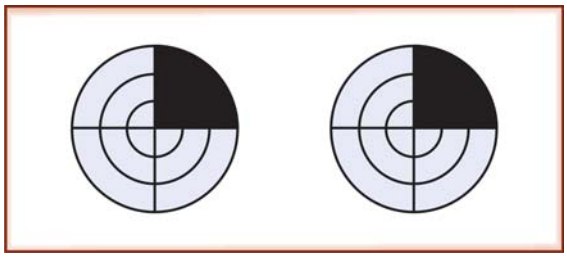

Fig. 20.8: Complete right superior homonymous quadrantanopia

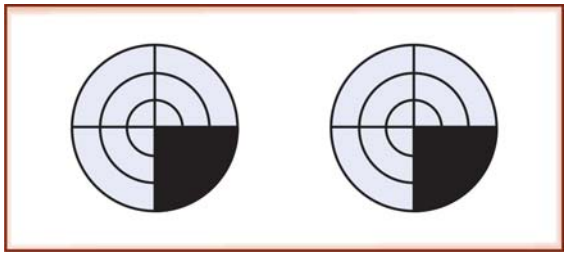

Fig. 20.9: Complete right inferior homonymous quadrantanopia

The lesions of the parietal lobe involve superior visual fibers of the optic radiations and produce contralateral inferior homonymous hemianopic defects or "pie on the floor" visual field defects (Fig. 20.9).

The macular fibers are spared owing to their widespread but segregated course in the optic radiations and their dual representation. The pupillary reactions remain unaffected and optic atrophy does not ensue.

Lesions of the Visual Cortex

The visual cortex is an area above and below the calcarine fissure which extends into the floor of the fissure as well as to the posterior pole of the occipital cortex. The lesions of the visual cortex classically produce homonymous hemianopic field defects (Fig. 20.10). They can be distinguished from the optic tract lesions by the absence of

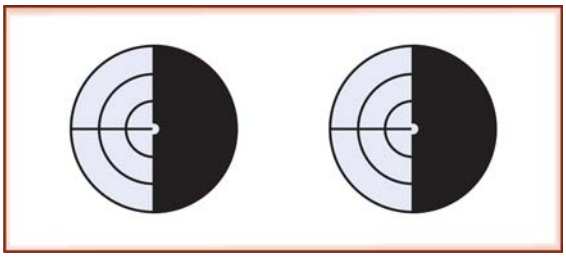

Fig. 20.10: Right homonymous hemianopia

abnormal pupillary reaction, congruity of the field defects and sparing of the macula.

The visual cortex is affected by injury, especially a fall on the back of the head or gunshot injury, cerebral tumors and cerebrovascular accidents. When both occipital lobes are damaged, complete blindness ensues. When the cortical lesion is situated near the angular gyrus, word-blindness is observed.

SYMPTOMATIC VISUAL DISTURBANCES

Amblyopia

Amblyopia is defined as unilateral or bilateral partial loss of sight without any organic ocular lesion. It usually develops in the first decade of life when the visual system is vulnerable to deprivation. Unilateral amblyopia is more common than bilateral. Common types of amblyopia, their causes and treatment are listed in Table 20.1.

Types of Amblyopia

1. *Strabismic amblyopia* is the most common type of amblyopia. The degree of deviation bears little relationship with the depth of amblyopia. Impairment of vision does not occur in fixing eye or in alternators. The correction of strabismus in early childhood prevents the occurrence of amblyopia.

Table 20.1: Causes and treatment of amblyopia

Types of amblyopia	*Causes*	*Treatment*
Strabismic	Suppression of deviating eye	Occlusion therapy
Anisometropic	Significant difference in the refractive errors of the two eyes	Correction of refractive error with glasses or contact lenses followed by occlusion therapy
Deprivation	Organic diseases such as ptosis, corneal opacity or cataract	Early surgery and occlusion therapy in suitable cases
Bilateral	Uncorrected bilateral hypermetropia	Correction of refractive error

2. *Anisometropic amblyopia* is found in patients whose refractive error in the two eyes differs by 2 D or more, and remains uncorrected for a long time. It is more commonly seen in unilateral hypermetropia or astigmatism than myopia, since the myopic eye is often brought in use for near work.
3. *Deprivation or amblyopia ex-anopsia* develops in early childhood owing to congenital or traumatic cataract, corneal opacity or developmental vitreoretinal disorders. The diseased eye does not receive visual stimulus (deprivation) and becomes lazy or dysfunctional. Therefore, early surgical intervention with restoration of clear media is indicated before the age of 2 years.
4. *Bilateral amblyopia* may occur in infants with uncorrected high hypermetropia. It may occasionally be found in young girls with psychosomatic disorders like hysteria.

Amaurosis

Strictly speaking, the term amaurosis should be used for a complete loss of vision without any organic change. Two types of amaurosis are commonly encountered in ophthalmic practice.

1. *Amaurosis fugax* is characterized by a sudden transient and painless loss of sight often due to transient circulatory failure. The attacks of blackout may be noticed in high altitude pilots, air travellers, as a prodromal symptom of central retinal artery or carotid artery occlusion, giant cell arteritis, hypertensive retinopathy, papilledema, Raynaud's disease and migraine.
2. *Uremic amaurosis* may occur in acute nephritis, renal failure and eclampsia. It results in a sudden bilateral blindness. The condition seems to be toxic in origin and is characterized by dilated, slow reacting pupils with normal optic disks. The recovery of vision may occur in 12 to 48 hours.

Loss of Vision

Loss of vision may be sudden or gradual, it may be painless or painful. Sudden loss of vision is considered as an emergency in ophthalmology and must be dealt with expeditiously and every attempt should be made to restore the lost vision.

Sudden Painless Loss of Vision

The sudden painless loss of vision may occur in a number of conditions listed in Table 20.2.

Gradual Painless Loss of Vision

Causes of gradual painless loss of vision differ in children and adults, and are listed in Table 20.3.

Table 20.2: Unilateral and bilateral causes of sudden painless loss of vision

Unilateral	*Bilateral*
Vitreous hemorrhage	Occipital lobe infarction
Retinal vascular occlusion	Diabetic retinopathy
Retinal detachment	Hypertensive retinopathy grade IV
Traumatic dislocation of lens	Posterior uveitis

Table 20.3: Causes of gradual painless visual impairment in children and adults

Children	*Adults*
Refractive error	Cataract
Developmental cataract	Glaucoma
Juvenile glaucoma	Corneal dystrophy
Keratoconus	Diabetic retinopathy
Hereditary macular dystrophy	Age-related macular degeneration

Sudden Painful Loss of Vision

The sudden painful loss of vision is more frequent than painless loss of vision and occurs in the following conditions.

1. Acute congestive stage of angle-closure glaucoma
2. Penetrating ocular injury
3. Acute uveitis
4. Ocular burns
5. Central corneal ulcer
6. Retrobulbar neuritis

Besides visual loss, symptomatic visual disturbances like black spots and flashes of light in front of eyes may occur. Some of the patients may complain of distortion in the shape of objects.

Black spots or *floaters* before eyes may be seen in vitreous degeneration due to myopia or old age, pars planitis, posterior uveitis, vitreous hemorrhage and retinal detachment. Occasionally black spots may be seen without any ocular pathology, the condition is known as *muscae volitantes*.

Flashes of light or *photopsia* may need special attention and investigations as they are prodromal symptoms of neuroretinitis, posterior vitreous detachment, retinal detachment and migraine.

Distortion of objects or *metamorphopsia* is an important symptom of macular lesion. It may occur in macular edema, central serous retinopathy, macular choroiditis, macular hole and macular pucker.

Migraine

Migraine is a disorder characterized by repetitive bouts of unilateral headache occurring more frequently in women than men.

Etiology

The etiology of migraine is not known. Heredity, hunger, psychic stress, pregnancy, menstruation, oral contraceptives and endocrine disorders have been considered as risk factors of migraine. Vasoconstrictive changes in the brain initiate the symptoms of migraine with neurological manifestations, while the subsequent vasodilatation results in hemicrania.

Clinical Features

Classical migraine is a clinical entity in which an aura of neurological disturbances, generally visual but occasionally motor or sensory, precedes the development of severe hemicrania. The visual aura presents a positive scotoma in the visual field which has a peculiar shimmering character. The scotoma gradually increases in size covering nearly one-half of the field and has bright spots as well as rays of various colors arranged in a zigzag manner. It is called *fortification spectra* (teichopsia) or *scintillating scotoma*. Despite its changing size and clouding of the field of vision, the fixation point is usually seen. Vision clears in about 15 to 20 minutes, but the aura is soon followed by a violent headache (hemicrania) accompanied with nausea, vomiting and giddiness. Depression and gastrointestinal symptoms may be associated with hemicrania. Migrainous attacks occur periodically and vary in severity. As the age advances, the scotoma may occur without headache or *vice versa.*

Ophthalmoplegic migraine is an uncommon disorder usually having its onset in the first decade of life. The attack of migraine is followed by partial paralysis of the third and/or the sixth cranial nerve. The paralysis may last for days or weeks. Ptosis, limitation of ocular movements, semidilated pupils and sluggish pupillary reactions are the classical features. Recovery is gradual and tends to become less complete with repeated attacks.

Treatment

The acute attack of migraine should be managed by administration of analgesics: aspirin, paracetamol, ibuprofen, etc. Ergotamine 1 mg with 100 mg caffeine and 4 mg dihydroergotamine may be given as a single dose. The drug is contraindicated in pregnancy, cardiovascular diseases and renal failure. As an alternative, sumatriptan 25 mg can be administered orally and repeated after 2 hours if pain is not relieved. Sumatriptan 6 mg can also be given subcutaneously as a single dose. Propranolol (10-80 mg/day upto a maximum dose of 160-240 mg/day), amitriptyline and calcium channel blockers (Flunarizine) may be taken on long-term basis for the prophylaxis of migraine.

Night-blindness

Poor or feeble vision in twilight or in night is known as *night-blindness* or *nyctalopia.* Night-blindness mainly occurs due to interference with functions of rods owing to deficiency in visual purple. Early night-blindness causes prolongation of dark adaptation time which can be detected by dark adaptometer.

The important causes of night-blindness are listed below.

1. Xerophthalmia
2. Primary pigmentary degeneration of retina
3. High myopia
4. Advanced open-angle glaucoma
5. Disseminated chorioretinal atrophy
6. Portal cirrhosis
7. Oguchi's disease, and
8. Congenital night-blindness.

Day-blindness

Poor vision in bright light or daylight is known as *day-blindness* or *hemeralopia.* It is mainly due to affection of cones at the macula. Central macular choroiditis, macular burn, retinochoroidal coloboma involving the macula and cone dystrophy cause day-blindness. Central corneal opacity and nuclear cataract lead to poor vision in bright light owing to constriction of pupil, but vision improves in dim light as the pupil dilates and the peripheral retina is used for vision.

Colored Vision (Chromatopsia)

Chromatopsia may occur in some cases of resolving optic neuritis. *Erythropsia* (red vision) occurs after cataract extraction when the eyes are exposed to sunlight. Objects appear red and the patient gets disturbed. Erythropsia may also develop in snow-blindness. Yellow vision (*xanthopsia*) occurs in jaundice, nuclear sclerosis and digitalis intoxication. Blue vision (*cyanopsia*) may occur for some time (a few days to months) after the removal of cataract and implantation of an intraocular lens (IOL). The natural lens reduces the amount of blue light reaching the retina. In patients with recent IOL implantation, more blue light than usual falls on the retina leading to bluish tinge. This invariably lasts transiently and ultimately gives way to normal color vision.

Color Blindness

An inability to identify colors suggests color blindness or *acromatopsia*. Color blindness may be congenital or acquired. The latter may be found in the affection of the retina and the choroid. The involvement of cones in these disorders affect mostly the blue end of the visible spectrum.

The cones mediate the color vision. There are three classes of cones in the human retina with different but overlapping spectral sensitivities. Color vision is dependent upon hue, saturation and brightness.

Complete color blindness is a rarity, however, partial or defective color perception is not uncommon. Color blindness is more common in males (3-4%) than in females (0.3%).

The defective color perception is a hereditary condition transmitted through females who usually remain unaffected (sex-linked). Most frequently, red and green colors are confused causing danger in certain occupations, particularly in railways and navigations.

Classification

Color blindness may be classified as:

1. Lack of color differentiation (achromatopsia), and
2. Color deficiency (dyschromatopsia).

Achromatopsia is a condition in which sensations of color are absent and vision is monochromatic. It is often associated with nystagmus, photophobia and poor visual acuity. The patient does not perceive any color, all colors appear gray and of different brightness.

Dyschromatopsia is a condition wherein color confusion occurs. Dyschromates have a bivalent color system rather than the trivalent. The defect is probably due to the absence of one of the photopigments normally present in the foveal cones. The dichromates are divided into three groups.

1. *Protanopes* are insensitive to red light (defective red sensation). They confuse red, yellow and green.
2. *Deuteranopes* have a defective green sensation but can match all colors with red and blue.

3. *Tritanopes* are very rare. They have some insensitivity to blue light but can match all colors with red and green.

More frequently, in dichromates the defects are milder. Depending on the spectral location of differences in color matching, they are classified as having *protanomaly*, *deuteranomaly* and *tritanomaly* or the subjects are known as *protans* (insensitive to deep red), *deutrans* (insensitive to light green) and *tritans* (insensitive to blue-green).

The various tests for determining color blindness have already been described in the chapter on *Examination of the Eye*.

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CHAPTER

21 Intraocular Tumors

Tumors arising from the uveal tract and the retina are described under intraocular tumors. They are usually malignant and may prove fatal.

TUMORS OF THE IRIS

Nevus of the Iris

Benign nevi of the iris are common. The nevus presents as a discrete, flat or elevated, lesion on the iris stroma. The iris nevi may be associated with neurofibromatosis and choroidal melanomas. Histologically, iris nevus appears as a collection of branching dendritic cells or spindle cells. Clinically, iris nevus may cause distortion of the pupil and ectropion uveae.

Malignant Melanoma of the Iris

Clinical Features

Malignant melanoma of the iris may present as a solitary pigmented or nonpigmented nodule usually located in the lower half of the iris. An ipsilateral hyperchromic heterochromia, ectropion of uveal pigment, distortion of the pupil, neovascularization of the iris, raised intraocular pressure and localized lenticular opacities support the diagnosis of malignant melanoma of the iris.

Differential Diagnosis

Iris melanoma must be differentiated from iris nevus, iris granuloma, leiomyoma, xanthogranuloma, iris cyst and secondaries in the iris.

Pathology

Histologically, most iris melanomas are slow growing and composed of spindle A or spindle B cells. They rarely metastasize and mortality is much lower than the melanomas of ciliary body and choroid.

Treatment

Malignant melanoma of the iris must not be dealt with radical excision, but should be periodically followed with meticulous clinical documentation. Rarely, broad iridectomy is needed for the tumor invading the angle. Diffuse melanoma of the iris warrants enucleation.

TUMORS OF THE CILIARY BODY

Malignant Melanoma of the Ciliary Body

Clinical Features

Malignant melanoma of the ciliary body often causes disturbance of vision due to distortion of the lens and interference with the action of ciliary muscle. The presence of conspicuous dilated one or two ciliary vessels (Fig. 21.1), and appearance of a dark crescent at the root of iris resembling iridodialysis, are characteristic signs of the tumor. The diagnosis may be confirmed on gonioscopy. The malignant melanoma of the ciliary body may be visible on oblique illumination in a dilated pupil

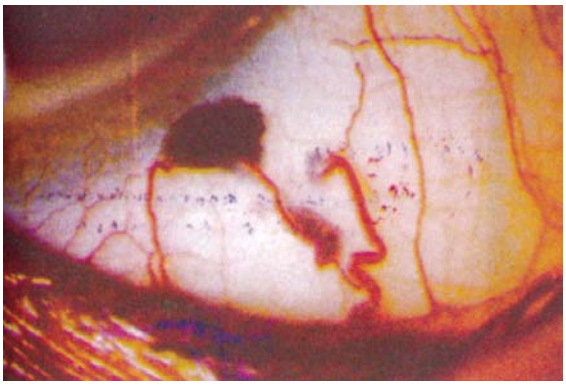


Fig. 21.1: Sentinel vessels in a case of malignant melanoma of ciliary body (*Courtesy*: Sankara Nethralaya, Chennai)

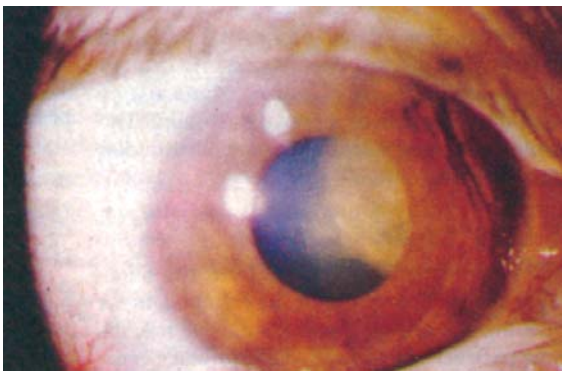


Fig. 21.2: Ciliary body melanoma through a dilated pupil (*Courtesy*: Sankara Nethralaya, Chennai)

(Fig. 21.2). The growth is often opaque to transillumination. Sometimes, the tumor is flat. Occasionally, the malignant melanoma of the ciliary body undergoes necrosis and causes anterior uveitis. The posterior extension of the tumor into the adjacent choroid can produce a nonrhegmatogenous retinal detachment which may involve the macula and cause impairment of vision.

Diagnosis

Ultrasound biomicroscopy is useful in the diagnosis of melanoma of the ciliary body. It can differentiate between a cyst and a tumor of the ciliary body (Fig. 21.3) as well as define the posterior extent of the tumor.

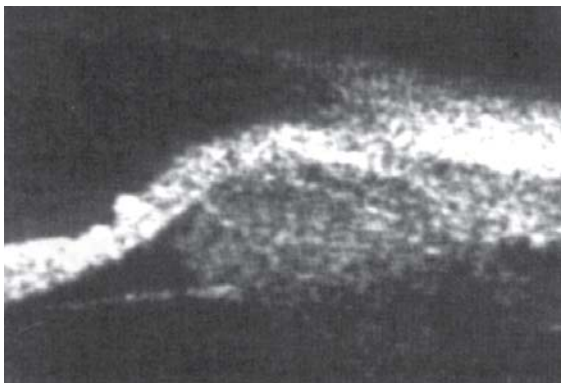


Fig. 21.3: Ultrasound biomicroscopy showing a small ciliary body tumor (*Courtesy*: Dr Muna Bhinde, Sankara Nethralaya, Chennai)

Treatment

A small localized melanoma of the ciliary body is removed by partial resection. An annular ciliary melanoma needs enucleation.

TUMORS OF THE CHOROID

Nevus

Majority of the nevi of the uveal tract occur in choroid. Drusen may be seen overlying the nevi. Occasionally, localized serous detachment of the retinal pigment epithelium or the neurosensory retina may develop. The choroidal nevi remain stationary for a long period, however, a few may give rise to melanomas.

Melanocytoma

Melanocytoma presents as a jet-black lesion that usually appears in the peripapillary region and is composed of plum polyhedral cells.

Hemangioma

Hemangiomas of the choroid occur in two forms: localized and diffuse.

Localized choroidal hemangioma is a red or orange colored tumor localized in the postequatorial region of the fundus. The involvement of macula results in blurred vision and metamorphopsia. The lesion affects the retinal pigment epithelium and causes cystoid macular degeneration (RPE).

Diffuse choroidal hemangioma is usually seen in patients with Sturge-Weber syndrome. It presents a reddish-orange fundus appearance that is referred as *tomato ketchup* fundus. The diffuse choroidal hemangioma can cause secondary glaucoma and exudative retinal detachment. It is treated with laser photocoagulation.

Malignant Melanoma of the Choroid

Malignant melanoma of the choroid is commonest (85%) among the uveal melanomas. It usually occurs between 40 and 60 years of age, and affects both sexes equally. It predominantly affects white races and has a predilection for the temporal half of the choroid. Nearly 10% of painful atrophic blind eyes contain unsuspected malignant melanomas.

Types

Melanomas of the choroid may occur in two forms: circumscribed type and diffuse type.

The *circumscribed melanoma* is almost always primary, single and unilateral. It is usually pigmented (Fig. 21.4), but unpigmented growth is not rare. Metastasis from the melanotic growth is often unpigmented.

The *diffuse melanoma* presents a slaty-gray pigmentation in the retina.

Pathology

Histologically, malignant melanoma can be divided into four cell types (*Callender's classification*): spindle cell melanoma (fascicular is an arrangement of spindle cells in a palisading or parallel rows), epithelioid cell melanoma, mixed cell melanoma and necrotic melanoma.

Clinical Features

Most malignant melanomas of the choroid have symptom-free onset. Visual impairment appears with the involvement of macula or with extension of retinal detachment.

The clinical course of the tumor is usually divided into four stages:

Quiescent stage The tumor generally arises from the outer layer of the choroid as a lens-shaped mass pushing the retina over it. Orange patches appear in the RPE due to the accumulation of lipofuscin. When the membrane of Bruch is ruptured, it assumes a collar-botton or mushroom-shaped configuration in the subretinal space (Fig. 21.5).

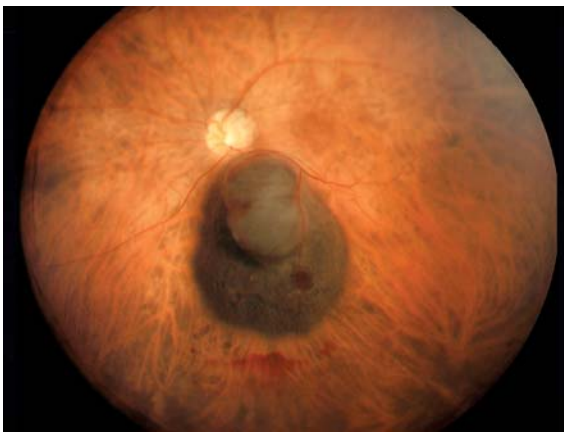


Fig. 21.4: Circumscribed malignant melanoma of choroid (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

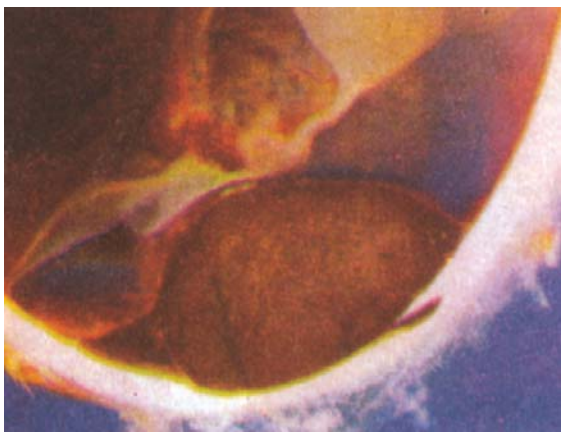


Fig. 21.5: Malignant melanoma of choroid on cross section (*Courtesy*: Sankara Nethralaya, Chennai)

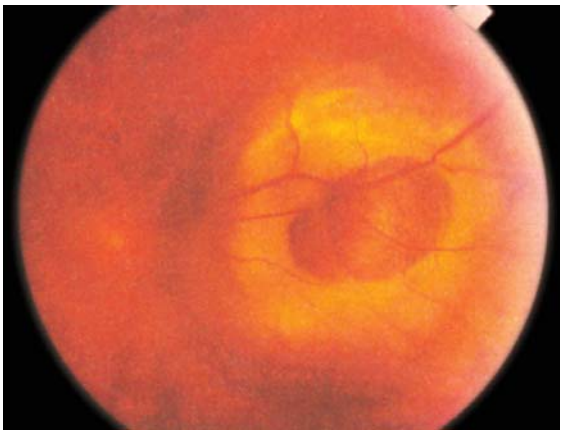


Fig. 21.6: Malignant melanoma of choroid with shallow retinal detachment (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

The malignant melanoma of choroid causes detachment of the retina around the tumor mass (Fig. 21.6) though the retina remains in contact with it at the summit. An abnormal slaty-gray or black pigmentation of the fundus is found in the diffuse infiltrating melanoma unassociated with retinal detachment.

Glaucomatous stage The glaucomatous stage is marked by severe pain and deterioration of vision due to rise of intraocular pressure (IOP). Secondary glaucoma may be due to the compression of vortex veins by the tumor mass or embarrassment of the drainage channels by the forward displacement of the lens-iris diaphragm. Direct invasion of the anterior chamber can also lead to glaucoma. Melanomalytic glaucoma may develop as a result of obstruction of the angle of the anterior chamber by melanin pigment following tumor necrosis.

Stage of extraocular extension The malignant melanoma of choroid may spread through scleral emissary channels to involve the bulbar surface of the orbit. It can also directly invade the underlying sclera and the overlying retina.

Stage of metastasis Metastasis almost invariably occurs from the malignant melanoma of choroid through hematogenous spread to the liver.

Differential Diagnosis

Malignant melanoma of the choroid must be differentiated from following diseases: (i) benign melanoma, (ii) hemangioma of the choroid, (iii) rhegmatogenous detachment of the retina, (iv) detachment of choroid, (v) parasitic cyst of choroid, and (vi) old choroidal hemorrhage.

Diagnosis

Majority of the choroidal melanomas can be diagnosed by indirect ophthalmoscopy, slit-lamp biomicroscopy with fundus contact lens, transillumination test, fluorescein angiography and ^{32}P uptake. B-scan ultrasonography is helpful in excluding rhegmatogenous retinal detachment especially when media are hazy.

Prognosis

Spindle cell melanomas have good prognosis (approximately 81%, 10 years survival) but epithelioid cell, mixed cell and necrotic melanomas have poor prognosis (less than 40%, 10 years survival). Diffuse melanomas also have poor prognosis.

Treatment

Malignant melanoma of the choroid may be managed on following guidelines:

1. An eye with large tumor without useful vision warrants enucleation, while conservative procedures should be applied in eyes with small melanomas and salvagable vision. The alternative modalities of management include periodic follow-up supported with stereoscopic colored fundus photographs.
2. Photocoagulation is advocated in malignant melanomas of less than 10 mm in width and 3 mm in elevation. The tumor should neither be located near the foveola nor be associated with subretinal fluid.
3. Irradiation of tumor can be performed by radioactive plaques comprising cobalt-60 or ruthenium-106.

4. In selective cases of peripherally located malignant melanoma a full-thickness local resection can be performed.
5. Cryopexy (–60°C) may be done as a primary procedure or preferably as a secondary procedure after the photocoagulation treatment.
6. Most patients with systemic metastasis need palliative radiation and chemotherapy.

RETINOBLASTOMA

Retinoblastoma is the most common primary intraocular malignancy of childhood. Nearly two-thirds of cases appear before the end of second year. There is no racial predisposition. A higher incidence of retinoblastoma is reported in males than females.

Genetics

The tumor is bilateral in 30-40% cases. The majority of unilateral retinoblastomas result from somatic mutations, but all bilateral cases are genetically determined by a tumor suppressor gene, RB1. Bilateral retinoblastoma represents a germinal mutation. It is associated with deletion of the q14 band of chromosome 13. In the absence of this antioncogenic chromosome, the retinal cell division continues unchecked causing retinoblastoma.

Most of the retinoblastomas are sporadic, while others occur in families. The tumor is transmitted as an autosomal dominant trait with irregular penetrance. Several families are known wherein three or four successive generations are affected.

Origin

The tumor arises from the premature cells of photoreceptor elements in the outer retinal layers as there is similarity between the fetal retinal cells and the rosette forming cells of retinoblastoma. Retinoblastoma is usually of multicentric origin.

Histopathology

Histologically, retinoblastomas are classified into two categories:
1. Differentiated, and
2. Undifferentiated.

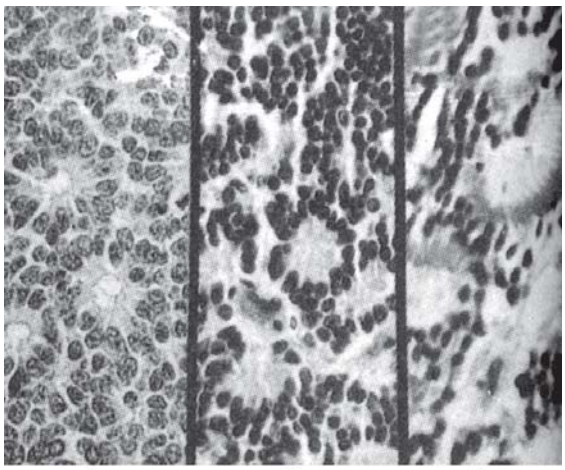
Differentiated Retinoblastoma

The tumor mass is composed of small closely packed round or polygonal cells with large darkly staining nuclei and scanty cytoplasm. The formation of *Flexner-Wintersteiner rosettes* (Fig. 21.7) is a strong evidence of differentiation. Less commonly *Homer-Wright rosettes* may be found.

Occasionally, well-differentiated retinoblastoma may have areas composed of *fleurettes*. Fleurettes are flower-like groupings of tumor cells within the retinoblastoma, and represent photoreceptor differentiation in the tumor.

Undifferentiated Retinoblastoma

The tumor consists mainly of anaplastic cells with hyperchromatic nuclei and scanty cytoplasm,



Figs 21.7A to C: Microphotograph showing: A. Flexner-Wintersteiner rosette, B. Homer-Wright rosette and C. Fleurettes (*Courtesy*: Drs J Biswas and M Shanmugam, Sankara Nethralaya, Chennai)

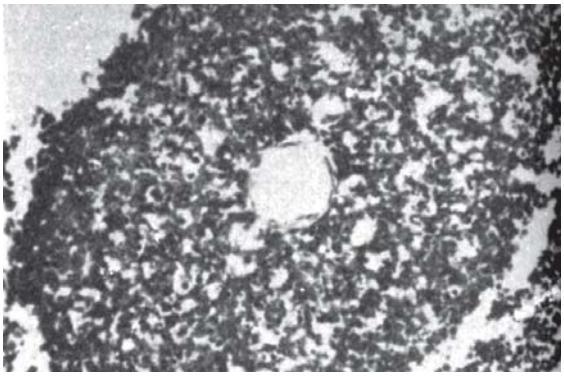


Fig. 21.8: Retinoblastoma—pseudorosette

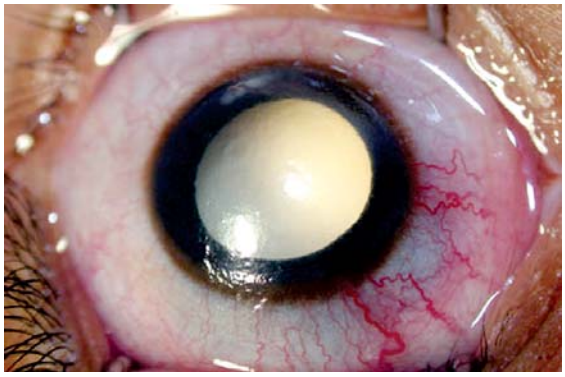


Fig. 21.9: Retinoblastoma presenting a cat's eye reflex (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

arranged in sheets and around the lumen of vessels. This arrangement is called *pseudorosettes* (Fig. 21.8). The mitotic figures are usually numerous and cell necrosis is widespread. Other histological features may include calcification and perivascular deposits of deoxyribonucleic acid.

Calcification is an important diagnostic feature of endophytic retinoblastoma. It occurs mainly in areas of necrosis. It can be detected by radiology and ultrasonography.

Clinical Features

Leukocoria or a white pupillary reflex is the most common (50%) presenting feature of retinoblastoma. Strabismus (esotropia) is the next common mode of presentation (about 20%).

Like malignant melanoma of the choroid, the clinical course of retinoblastoma is divided into four stages.

Stage of quiescence: The stage of quiescence may last from six months to a year and the child remains symptom-free, but the yellowish-white reflex, *amaurotic cat's eye reflex* (Fig. 21.9), in a child's eye may attract the attention of the parents. The pupil is usually dilated and nonreacting and the eye may be virtually blind.

Ophthalmoscopy may reveal the presence of a growth. Depending on the growth pattern and fundus appearance, the tumor may be classified into two clinical categories:

1. Endophytum—the tumor growing towards the vitreous, and
2. Exophytum—the tumor growing into the subretinal space.

Endophytic retinoblastoma protrudes from the retina into the vitreous. It is easily seen by an ophthalmoscope and there is no associated retinal detachment. It appears as a white fluffy or pink colored mass having neovascularization over the summit (Fig. 21.10)). The calcified mass may resemble a cottage cheese. There may be more than one growth in the same eye. Occasionally the endophytic retinoblastoma may simulate endophthalmitis.

Exophytic retinoblastoma grows in the subretinal space and causes extensive detachment of the retina. The growth may be masked when vitreous is hazy.

Stage of glaucoma: The child is irritable and cries with pain due to secondary glaucoma. The rise in intraocular pressure occurs as a result of distension of globe or blockage of the drainage channels at the angle of the anterior chamber. The conjunc-

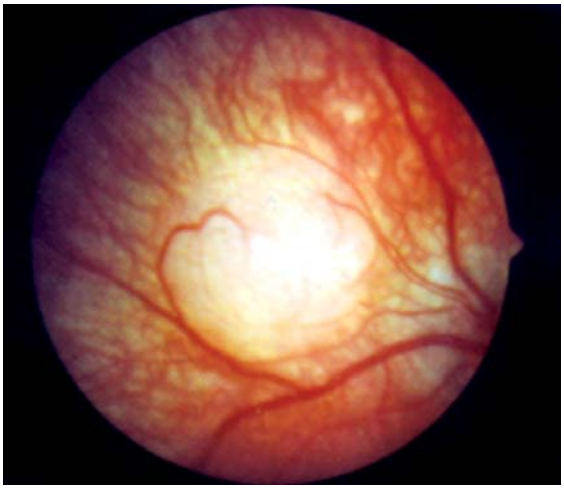


Fig. 21.10: Retinoblastoma in superotemporal quadrant (*Courtesy*: Prof. DM Robertson, Rochester)

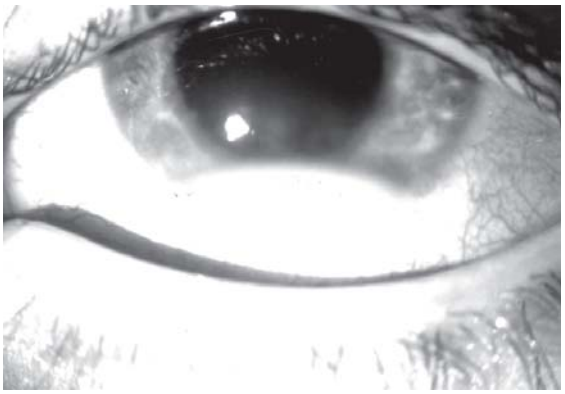


Fig. 21.11: Pseudohypopyon with convex level in retinoblastoma (*Courtesy*: Prof. DM Robertson, Rochester)

tiva is injected and the eyeball is enlarged. Retinoblastoma cells may enter the anterior chamber and give rise to iris nodules or pseudohypopyon. The hypopyon has a convex level (Fig. 21.11). The clinical picture mimics anterior uveitis. Secondary glaucoma and rubeosis iridis occur frequently.

Stage of extraocular extension: Extraocular extension of the tumor occurs by local invasion of the orbital tissue through scleral emissaries or by direct extension in the optic nerve. The extraocular fungating mass projects between the lids and undergoes secondary infection mimicking panophthalmitis. Enlargement of preauricular and cervical lymph nodes is common.

Stage of metastasis: The common identified sites of metastatic spread of retinoblastoma include brain, flat bones of cranium, iliac crest and sternum, and lymph nodes. Intracranial involvement is attributed to a direct extension of the growth along the optic nerve. Blood-borne metastasis is relatively uncommon.

Associated Conditions

Retinocytoma or *retinoma* is a benign counterpart of retinoblastoma or a type of retinoblastoma which has undergone differentiation. However, retinocytoma differs histologically from retinoblastoma on the following points: (i) retinocytoma cells have more cytoplasm and show no mitosis, and (ii) necrosis is usually absent in retinocytoma.

When bilateral retinoblastoma is associated with a tumor of pineal gland (pinealoblastoma), the condition is called *trilateral retinoblastoma*.

Diagnosis

The diagnosis of retinoblastoma requires a careful examination of the child under general anesthesia. The examination should include measurement of corneal diameter and IOP, detailed fundus examination under full mydriasis to assess the extent of the tumor and examination of the fellow eye to rule out bilateral involvement.

Radiographs of orbit may show intraocular and intracranial calcifications and enlargement of the optic foramen. Ultrasonography and CT scanning can be helpful in demonstrating the characteristic calcific densities within a retinoblastoma. B-scan ultrasonography may show a cauliflower-like mass arising from the retina

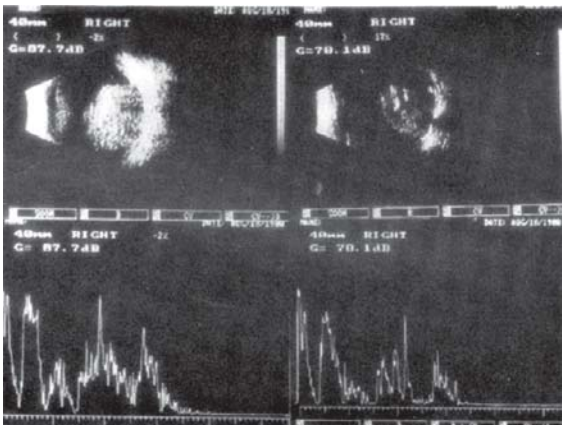


Fig. 21.12: B-scan ultrasonography showing an irregularly large echogenic mass in vitreous arising from retina (*Courtesy*: Dr TP Das, LVPEI, Hyderabad)

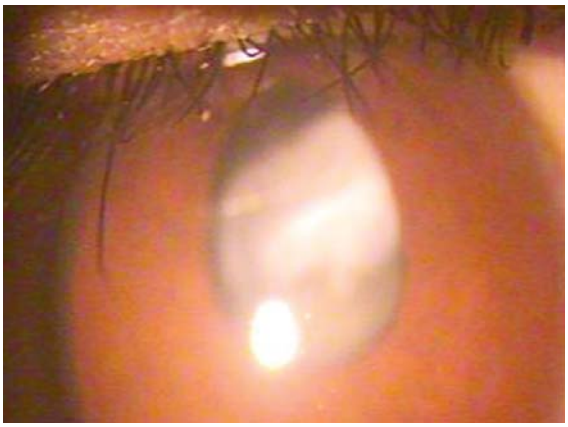


Fig. 21.13: Pseudoglioma

(Fig. 21.12) and numerous high intensity internal echoes throughout the mass lesion and orbital shadowing due to calcification. CT scan can determine the extent of tumor invasion in the optic nerve, orbit, and central nervous system (CNS).

Lumbar puncture and bone marrow aspiration may be needed in selected patients of retinoblastoma with metastasis. Other diagnostic modalities include estimation of aqueous lactic acid dehydrogenase level and fine needle aspiration or excisional biopsy. Magnetic resonance imaging is useful to detect the extraocular extension of retinoblastoma.

Differential Diagnosis

A number of conditions may mimic the clinical picture of retinoblastoma and they cause a diagnostic puzzle. All of them are non-neoplastic and grouped together under *pseudoglioma* (Fig. 21.13) and are listed below.

1. Congenital cataract
2. Acute iridocyclitis with vitreous exudation
3. Persistent hyperplastic primary vitreous
4. Toxocariasis
5. Retinal detachment
6. Retinopathy of prematurity
7. Coat's disease
8. Retinal astrocytoma.

Treatment

Current treatment modalities available for retinoblastoma include enucleation, chemoreduction with local tumor ablation, photocoagulation, cryotherapy, external beam radiation and brachytherapy.

Enucleation: It is indicated when retinoblastoma involves more than 50% of globe or when orbital or optic nerve involvement is suspected. The optic nerve stump must be of more than 10 mm avoiding the potential for instrumental globe penetration. Histopathological confirmation of the involvement of optic nerve warrants postoperative radiation or chemotherapy.

Debulking of the orbit: The debulking is indicated in the stage of extraocular extension. It should be followed by chemotherapy and radiation.

Chemoreduction with local tumor ablation: It is a significant advancement in the management of retinoblastoma. Intravenous administration of chemotherapeutic drugs such as carboplatin, vincristine, etoposide and cyclosporine every three weeks for 4 to 9 cycles can reduce the size of the tumor (Fig. 21.14). It is followed by transpupillary laser ablation. Systemic chemotherapy is indicated for children with metastatic disease.

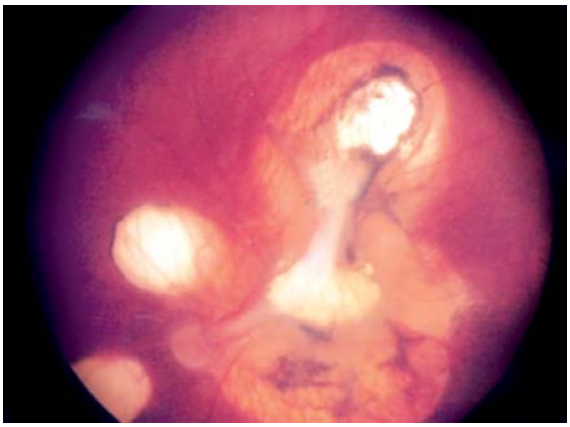


Fig. 21.14: Regression of retinoblastoma after chemotherapy and photocoagulation(*Courtesy*: Prof. DM Robertson, Rochester)

Photocoagulation: When retinoblastoma is smaller than 3 mm in apical height and less than 10 mm in basal diameter, a direct photocoagulation treatment is done over the entire tumor.

Cryotherapy: The small (3 mm × 10 mm) anteriorly located tumors are managed by cryotherapy which is applied under direct vision with a triple freeze-thaw technique.

External beam radiation therapy: Retinoblastomas are sensitive to radiation. Radiation (4000-4500 cGY) is delivered over a period of 4-6 weeks in children with bilateral tumors. The technique has two major disadvantages: (i) radiation-related sequelae such as cataract and optic neuropathy may develop, and (ii) there is a high risk of development of second primary malignancy (osteosarcoma).

Bachytherapy or *plaque radiotherapy*: Small size retinoblastoma (8 mm × 16 mm) can be managed by iodine-125 plaque radiotherapy. The technique can cause radiation optic neuropathy or vasculopathy.

Spontaneous Regression

Retinoblastoma can undergo spontaneous regression following a complete necrosis. The mechanism is not well understood. The eye becomes phthisical and filled with islands of calcified cells within a mass of fibrous tissue.

Prognosis

Advances in the diagnosis and management of retinoblastoma have significantly improved the prognosis. Children treated early and adequately have a survival rate of over 95%. Undifferentiated tumors with optic nerve involvement or massive choroidal invasion have a poor prognosis. Bilateral retinoblastomas with CNS involvement have very poor prognosis.

Second Tumor

The retinoblastoma survivors are prone to develop a second primary neoplasm (2-15%) irrespective of their treatment modality. All patients of retinoblastoma must be followed at regular intervals—six weekly for six months and six monthly for three years. The chances of recurrence after three years are remote. The near relatives of the patients of retinoblastoma should also be followed up closely.

METASTATIC TUMORS

Metastatic tumors occur commonly in adults and frequently involve the choroid.

The common primary tumors metastasizing to the eye are carcinoma of the breast in females (68%) and carcinoma of the lung (Fig. 21.15) in males (40%). Other primary tumors metastasizing to the eye arise from the gastrointestinal system, kidneys, prostate and skin. The primary site of approximately 25% of the tumors remain unknown.

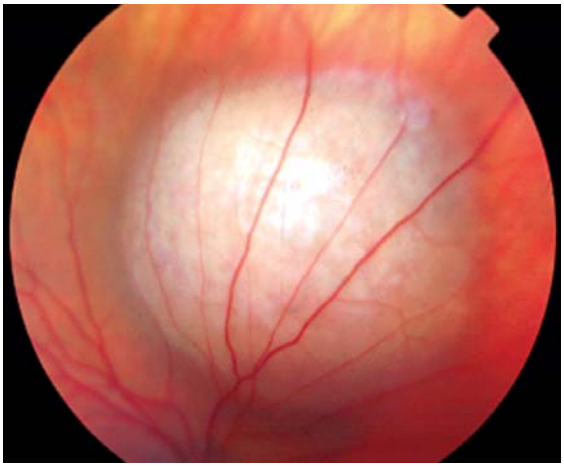


Fig. 21.15: Metastasis from carcinoma of lung manifesting as a yellow-white choroidal mass (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

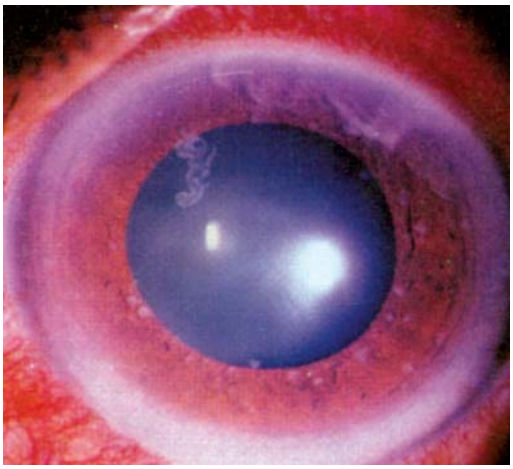


Fig. 21.16: Matastasis in iris presenting as anterior uveitis with fluffy exudates over iris (*Courtesy*: Dr J Biswas, Sankara Nethralaya, Chennai)

Metastasis to the eye usually occurs through hematogenous dissemination of the tumor cells. The choroid is most commonly involved due to its rich blood supply. The retina and the optic nerve are rarely involved. Histologically, the tumor may resemble with the primary lesion or may be less differentiated.

Metastasis in the iris and the ciliary body manifests as gray-white gelatinous nodules or appear as anterior uveitis (Fig. 21.16), rubeosis iridis or secondary glaucoma.

Patients with metastatic tumors of the choroid often complain of photophobia and impairment of vision. The choroidal lesions are bilateral, multiple, typically flat and look gray-yellow or yellow-white.

Metastasis in the retina, although rare, presents as cotton-wool spots near the blood vessels or may cause nonrhegmatogenous retinal detachment. The involvement of the optic nerve leads to papilledema.

Treatment

Chemotherapy is the frontline treatment modality especially for secondaries from the breast cancer. Rapid improvement may be obtained with radiotherapy (3500-5000 cGy) in most of the metastatic tumors. In spite of treatment, the prognosis is poor.

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CHAPTER

22 Injury to the Eye

OCULAR INJURIES

In spite of the protection given by the bony orbit, the eyelids and the eyelashes and their movements during blink reflex, injuries to the eye are not uncommon. The nature of injury differs according to the occupation like severe gun-shot wounds or blast injuries occur in military combat. Foreign bodies or flying objects may hit and lodge upon the conjunctiva or the cornea or enter the eye in the industrial workers. Children often sustain eye injuries during play due to bow and arrow, catapult, air-rifle and fireworks.

Classification

Depending on the nature of trauma the ocular injuries may be classified as:

1. Mechanical injuries:
   a. Extraocular foreign body
   b. Nonpenetrating injury (Blunt trauma)
   c. Penetrating injury
   d. Impalement injury
2. Chemical injuries:
   a. Alkali burn
   b. Acid burn
3. Injuries due to physical agent:
   a. Thermal
   b. Electrical
   c. Radiational
4. Indirect ocular trauma.

EXTRAOCULAR FOREIGN BODIES

Etiology

Foreign bodies such as small particles of coal, dust, wood, stone, husk of paddy, wings of insects and iron may hit upon the surface of the eyeball and may be retained either in the sulcus subtarsalis or on the cornea. Minute corneal foreign bodies are generally washed away by tear flow.

Clinical Features

A retained foreign body (FB) in the palpebral conjunctiva causes much irritation and watering. Chips of metal (iron or steel) penetrate into the epithelium (Fig. 22.1) or stroma of the cornea, and cause pain and photophobia.

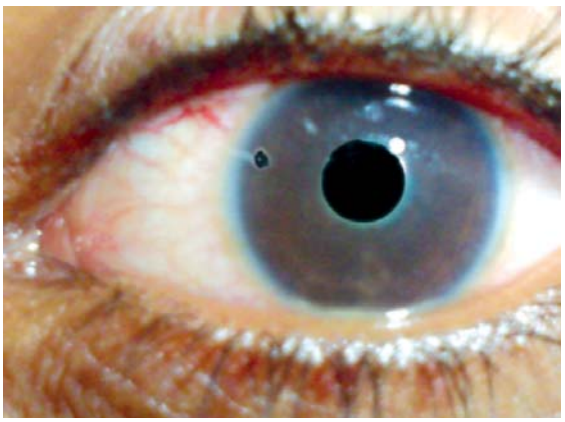

Fig. 22.1: Foreign body on cornea

Complications

An infected foreign body can cause conjunctivitis while a retained corneal foreign body leads to corneal ulcer. Corneal opacities and rust-ring (iron FB) are common sequelae.

Diagnosis

Small foreign bodies are detected with the help of a loupe or on slit-lamp examination. When a foreign body is not found, a careful examination may reveal the presence of an abrasion caused by the foreign body.

Prophylaxis

The wearing of safety goggles must be made compulsory for the workers who are engaged in grinding and lathe industries.

Treatment

A foreign body found on the cornea or the conjunctiva must be removed as soon as possible. Most particles of coal and dust can be removed with the help of a cotton swab after surface anesthesia, but embedded ones need a sharp needle. The iron particles when lodged in the deeper part of the cornea should be removed by a forceps or a magnet in the operation theater. After the removal of the foreign body, the eye should be treated on the lines of a corneal ulcer.

NONPENETRATING INJURIES (BLUNT TRAUMA)

Nonpenetrating injuries of the eye are caused by blunt objects such as a blow or a tennis or a cricket ball. The result of a blunt trauma depends largely on the force of the impact. The injury may cause displacement, rupture and tear of the intraocular structures with or without rupture of the globe. It may cause an immediate serious diminution of vision or may have a delayed effect.

Ocular tissues are severely traumatized by a direct force impinging upon the cornea in blunt trauma which is transmitted through the aqueous and the vitreous backwards (compression wave force) as well as by an indirect rebounding compression wave force from the back of the eye. The globe often ruptures when it is violently banged against the orbital wall (indirect force).

Cornea

Minor corneal abrasions or extensive damage to the cornea are not uncommon. The corneal abrasions are very painful and can be recognized by fluorescein staining of the cornea (Fig. 22.2). Recurrent erosions of cornea are common following finger-nail injury. They may not respond to routine treatment; carbolic acid or trichloracetic acid cautery may be beneficial. A deep corneal opacity may develop as a result of stromal edema or due to rupture of Descemet's membrane. Rupture of the cornea, though rare, must be repaired immediately.

Anterior Chamber

Hyphema or blood in the anterior chamber (Fig. 22.3) frequently occurs from a blunt trauma. If it is massive (Fig. 22.4) or recurrent and

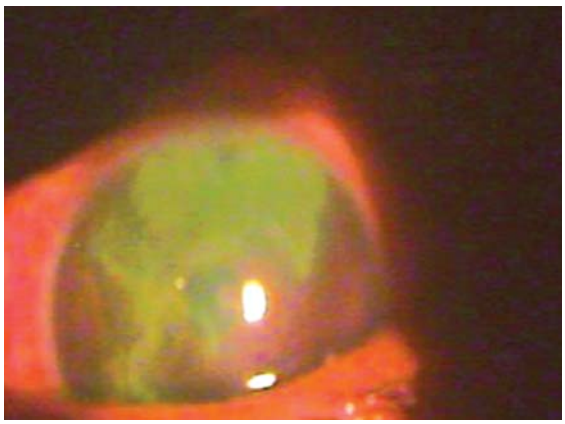

Fig. 22.2: Traumatic corneal ulcer (stained with fluorescein) and hyphema

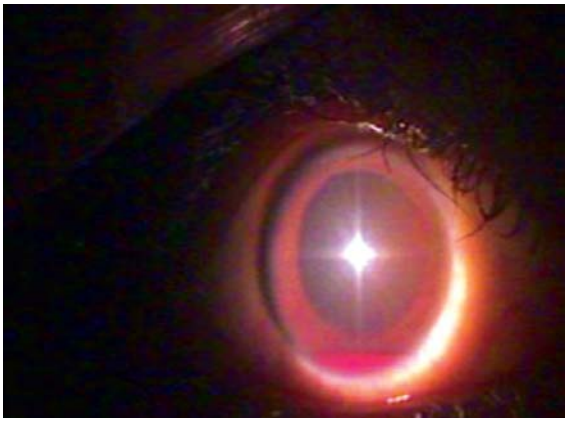

Fig. 22.3: Hyphema

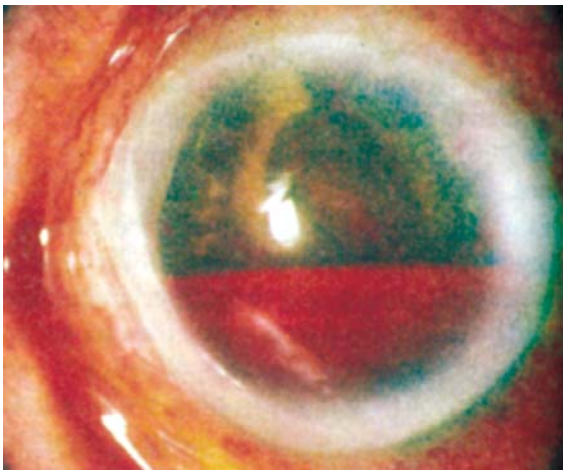

Fig. 22.4: Massive recurrent hyphema

accompanied by a rise of ocular tension, blood-staining of the cornea may ensue. The condition simulates dislocation of the lens in the anterior chamber. Lowering of the intraocular pressure and paracentesis of the anterior chamber (to evacuate the blood) prevent the blood-staining of the cornea.

Sclera

Rupture of the sclera occurs near the canal of Schlemm following a blunt injury, the tear is more or less concentric with the limbus and extends outwards and backwards. There may be prolapse of iris, recession of the angle of the anterior chamber and intraocular hemorrhage. In severe cases, the lens is expelled from the eye and the vitreous presents in the wound. There may be detachment of the retina with subretinal and suprachoroidal hemorrhages. The damage to ocular tissues is so extensive that the eye usually shrinks. The overlying conjunctiva may remain intact.

The wound should be carefully examined after incising the conjunctiva and sutured after reposing or excising the prolapsed iris.

Pupil, Iris and Ciliary Body

Traumatic miosis, mydriasis, irregular pupil, iridodialysis or aniridia and anteflexion or retroflexion of the iris may be seen after blunt trauma.

A *traumatic miosis* is due to the irritation of parasympathetics and may be associated with spasm of accommodation. *Traumatic mydriasis* may be transient or permanent. It occurs due to paralysis of parasympathetic nerve fibers. Rupture of the pupillary margin (sphincter tear) causes irregular and dilated pupil.

Iridodialysis occurs as a result of detachment of the iris from its ciliary attachment (Fig. 22.5). Iridodialysis may be complete (aniridia) and the

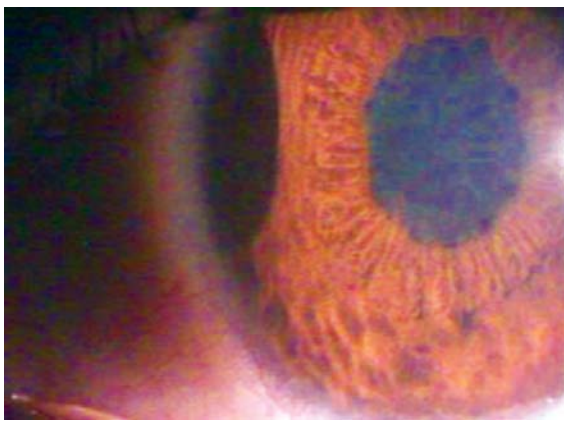

Fig. 22.5: Traumatic iridodialysis

detached iris sinks to the bottom of the anterior chamber. *Anteflexion* and *retroflexion of iris* are rare conditions often associated with extensive iridodialysis.The rupture of the ciliary body may sometimes occur. Traumatic uveitis may develop following a blunt trauma.

Cold compresses and cycloplegic eye drop should be employed. Suturing of the torn periphery of the iris and ruptured globe must be undertaken immediately.

Lens

Vossius ring, concussion cataract and subluxation or dislocation of the lens may occur following a blunt trauma.

Vossius ring is due to an impact of the iris onto the anterior surface of the lens produced by the force of a blunt injury. It consists of multiple brown granules of iris pigments arranged in a circular manner coinciding with the diameter of the constricted pupil.

Traumatic cataract develops partly due to mechanical injury to the lens fibers and largely due to damage to the lens capsule. The lens capsule ruptures usually at the posterior pole where it is thinnest. The aqueous humor permeates through the damaged capsule and causes opacification of the lens. The typical concussion cataract has a rosette shape and is situated in the posterior cortex (Fig. 22.6), and sometimes in the anterior cortex. The entrance of aqueous into the posterior cortex delineates the cortical sutures and from them the opacities radiate outwards in a feathery manner. The cataract may remain stationary or progress to total opacification of the lens. Rarely, a late rosette-shaped cataract may develop one or two years after the concussion injury.

Subluxation of the lens is due to a partial rupture of the zonule. Uniocular diplopia, irregular depth of the anterior chamber and segmental iridodonesis are the characteristic signs of subluxation of the lens. Ophthalmoscopy shows dual images

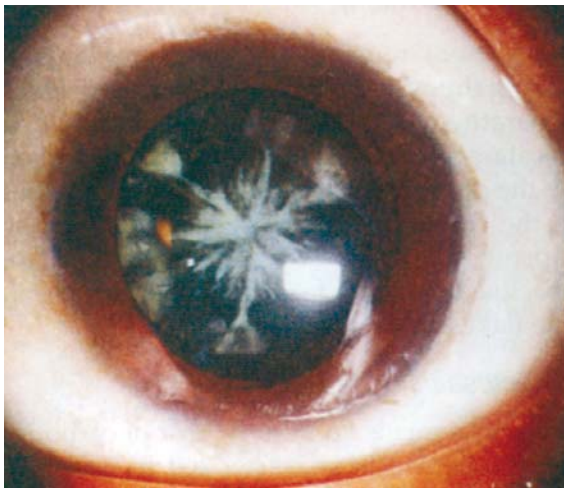

Fig. 22.6: Rosette-shaped cataract due to concussion

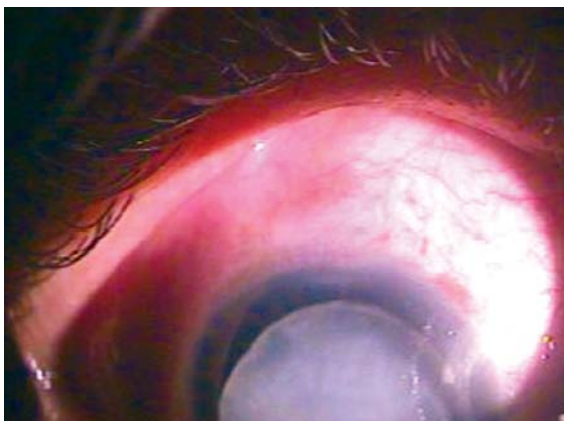

Fig. 22.7: Dislocation of cataractous lens in anterior chamber

of the optic disk through the phakic and the aphakic portion of the pupil.

Dislocation of the lens develops when the tear of the zonule is complete, the lens is dislocated posteriorly into the vitreous or anteriorly in the anterior chamber (Fig. 22.7). It may get dislocated in the subconjunctival space due to rupture of the sclera.

A clear lens in the anterior chamber is globular in shape and appears as an oil-globule. It can cause anterior uveitis and secondary glaucoma

(phacotopic glaucoma), hence warrants extraction. The lens in the vitreous may remain clear or turn opaque.

Traumatic cataracts are managed on general lines. Subluxated lens need not be removed unless it is opaque or causing secondary glaucoma.

Intraocular Pressure

In trivial trauma, after initial fluctuation, the IOP returns back to normal. A traumatic glaucoma may develop due to intense vasodilatation, iridocyclitis, recession of the angle of anterior chamber, dislocation of the lens and intraocular hemorrhage.

Rupture of the globe, with or without extrusion of the intraocular contents, and traumatic atrophy of the ciliary body cause ocular hypotonia.

Vitreous

Liquefaction of the vitreous associated with pigmentary vitreous opacities is common following a blunt trauma. Vitreous hemorrhage and retinitis proliferans are not uncommon after a concussion injury. Partial or complete vitreous detachment may occur. The partial detachment of the vitreous can produce a traction on the retina.

Choroid

Choroidal hemorrhage and rupture may follow a contusion injury. Small scattered hemorrhages in the choroid are common which subsequently lead to chorioretinal atrophy. The rupture of the choroid appears as a curved white concentric streak (Fig. 22.8) often on the temporal side of the disk over which the retinal vessels course. The edges of the streak are soon invaded by pigments. Occasionally, there may be multiple ruptures. A tear in the macular area abolishes the central vision.

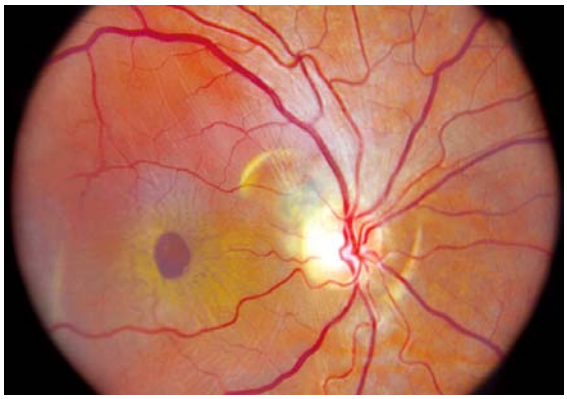

Fig. 22.8: Rupture of choroid and macular hole

Retina

Significant changes in retina may occur following a blunt trauma.

Commotio retinae or Berlin's edema usually develops after a blow on the eye. It is characterized by the presence of a cherry-red spot surrounded by a milky-white cloudiness (edema) of the macular area associated with the diminution of central vision. The condition may simulate central retinal artery occlusion. Later, the edema subsides and pigmentary deposits appear in the macula.

Macular cyst and *macular hole* may develop after a blunt ocular injury. A macular cyst on rupture produces a macular hole (Fig. 22.8). Both macular cyst and hole appear as a round red spot, but the hole has a punched-out appearance.

Retinal tear and *retinal detachment* may occur even after a minor trauma in myopic eyes.

Rupture of the retinal vessels causes *retinal hemorrhages* as well as *vitreous hemorrhage*. The organized vitreous hemorrhage may produce dense traction bands which on contraction detach the retina. A traumatic chorioretinitis is not infrequent following a blunt ocular trauma.

Fig. 22.9: Massive subconjunctival hemorrhage with hyphema

Optic Nerve

Injury to the optic nerve is frequent in the fracture of the base of skull. Avulsion of the nerve is rare but occurs in gun-shot injury. It is characterized by an excavated papilla, peripapillary hemorrhages and marked visual loss.

Blunt Trauma to the Ocular Adnexa

Lids and Conjunctiva

Contusion injuries to the lids lead to their enormous swelling and ecchymosis. It is often associated with subcutaneous extravasation of the blood (*black eye*) and subconjunctival hemorrhage (Fig. 22.9). A traumatic coloboma of the lid is not rare. The vertical wounds of the lids usually gape, while lacerated wounds leave ugly scars and deformities.

Cold compresses relieve ecchymosis caused by contusion. Vertical wounds of the lids must be sutured in layers, while irregular wounds may require plastic repair. Local and systemic antibiotics should be administered to control infection.

Lacrimal Canaliculi and Lacrimal Sac

Injury at the medial canthus may implicate the lacrimal canaliculi, sac and nasolacrimal duct and results in epiphora. It is often found in conjunction with craniofacial trauma in road traffic accidents. They need plastic repair. Dacryocystitis is a common sequela of injury to the lacrimal passage.

BLOW-OUT FRACTURE OF THE ORBIT

Etiology

The blow-out fracture of the orbit results from a blunt trauma to the orbit such as by a fist, cricket ball or football. The striking object produces an increased intraorbital pressure that is transmitted by bones to the weakest points, thereby shattering them (*hydraulic theory*). The *buckling theory* contends that the posterior transmission of a compressive force at the inferior orbital rim leads directly to a buckling of the orbital floor. The orbital bones break at their weakest sites, the orbital floor and the medial wall.

Clinical Features

An orbital blow-out fracture presents following features:

1. *Ecchymosis* of the eyelid and *emphysema* of the eyelid and the orbit.
2. *Diplopia* on the upward and the downward gaze, and both horizontal and vertical ocular movements are restricted. Restricted and painful vertical movements of the globe and diplopia suggest entrapment of the inferior rectus muscle. The entrapment can be confirmed on a *forced duction test*. Under topical anesthesia the eye is grasped at the limbus and rotated in the deficient direction of gaze. Limitation of passive movements of the eye confirms a restrictive etiology (extraocular muscle entrapment).
3. *Enophthalmos* (Fig. 22.10) occurs due to the prolapse of orbital tissues in the maxillary and ethmoid sinuses. Enophthalmos becomes significant as the orbital edema subsides.
4. *Hypoesthesia* in the the distribution of infraorbital nerve.

Fig. 22.10: Fracture floor of right orbit: showing enophthalmos and restricted elevation (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

Diagnosis

The diagnosis of a blow-out fracture is confirmed by obtaining both axial (Fig. 22.11) and coronal views of the orbit on CT scan. It is a better imaging modality for bony structures of the orbit than plain X-ray or MRI.

Treatment

Majority of the orbital fractures are treated on conservative lines but surgical intervention is indicated when: (i) fracture is large, (ii) diplopia persists for more than 10 days after the injury, and (iii) enophthalmos exceeds 2 mm.

The surgical approach through the lower eyelid includes: (i) elevation of periorbita from the orbital floor, (ii) release of entrapped inferior rectus muscle and orbital tissue, and (iii) placement of an implant.

Fracture Base of Skull involving Optic Nerve

The fracture of the base of skull implicates optic foramen and may cause optic atrophy or pulsating exophthalmos. The fracture is characterized by a wound at the lateral part of the eyebrow, loss of direct ipsilateral pupillary reaction and hemianopic field defects. The patient may suffer from

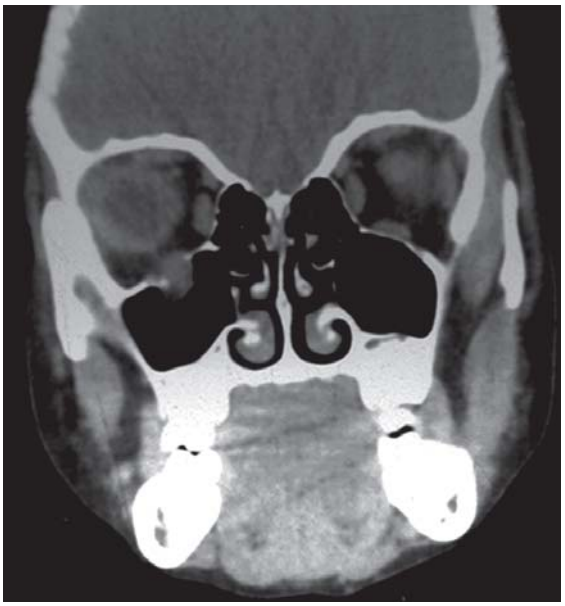

Fig. 22.11: CT scan showing fracture right floor of orbit (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

epistaxis and varying periods of unconsciousness. The pallor of the optic disk may be noticed 2-3 weeks after the injury. CT scan is a standard imaging technique for the diagnosis of base of skull fracture.

IMPALEMENT INJURIES

Impalement injuries to the orbit may occur when a child falls on a pencil held in his hand or by an arrow or a knife. The injury causes severe bleeding and orbital hematoma. The bleeding should be controlled, and the treatment is aimed to minimize the risk of orbital cellulitis.

PENETRATING INJURIES

The penetrating injuries of the eyeball are caused by sharp pointed objects: needle, knife, scissors or flying metallic foreign bodies. The injuries not only produce an immediate damage to the eye structures but may also introduce severe pyogenic infection into the eye or induce sympathetic ophthalmitis in the sound

eye. Patients with penetrating injuries of the eye may present with or without retention of a foreign body.

A sharp instrument or object may cause a linear or lacerated wound of lids, conjunctiva, cornea, sclera, iris, ciliary body and lens.

Wound of the Conjunctiva

The wound of the conjunctiva are often associated with a subconjunctival hemorrhage. Large wounds of the conjunctiva need repair otherwise symblepharon may develop.

Wounds of the Cornea

Corneal wounds are frequent and vary from a minor cut to an extensive one involving the sclera. They need repair (Fig. 22.12). A small wound of the cornea may heal quickly if treated on the lines of a corneal ulcer. It leaves a dense corneal opacity. A deep corneal wound is often associated with an iris prolapse (Fig. 22.13). The prolapsed iris must be abscised and the wound is sutured.

Wounds of the Sclera

The extent of the scleral wound should always be assessed after reflection of the conjunctiva. The incarcerated tissue, the iris, the lens and the vitreous, should be abscised and the sclera is sutured. When injury is extensive and panophthalmitis supervenes, repair of the wound is of no avail and the eye should then be eviscerated.

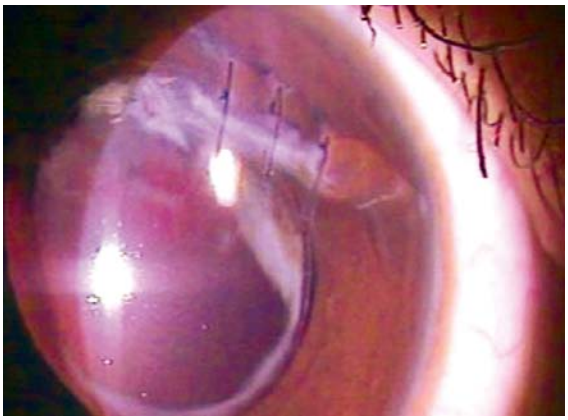

Fig. 22.12: Sutured corneal wound following penetrating injury

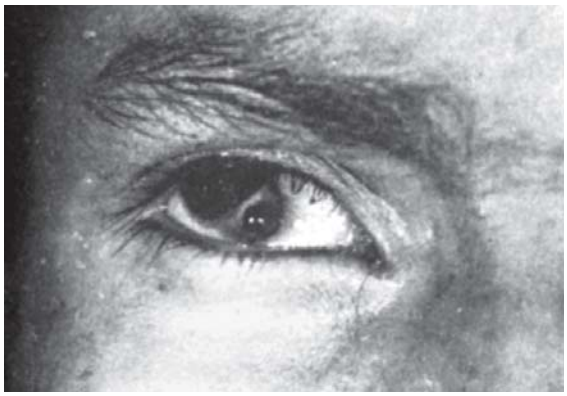

Fig. 22.13: Iris prolapse

Fig. 22.14: A linear penetrating injury of cornea with uveitis and cataract

Wounds of the Lens

Sharp instruments, especially the needles, perforate the capsule of the lens and the entry of aqueous produces a rosette-shaped lenticular opacity. If the wound is large, the lens soon becomes opaque (Fig. 22.14).

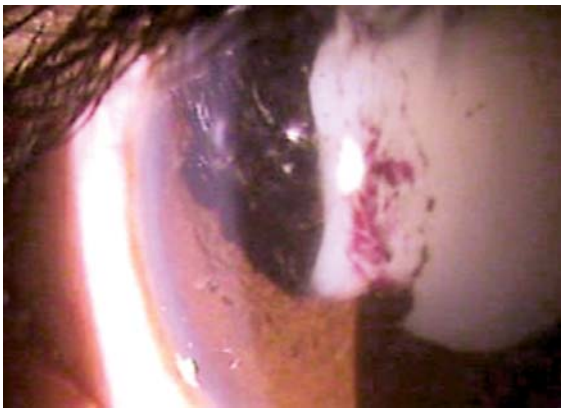

Fig. 22.15: Aphakia with thick lens matter following penetrating injury

In young individuals, the lens matter gets dissolved by the aqueous resulting in aphakia (Fig. 22.15), but in adults the hard nucleus persists.

Sometimes, the lens matter in the anterior chamber may induce secondary glaucoma or iridocyclitis. In such cases aspiration of lens matter is often necessary. Topical corticosteroids, cycloplegic and antibiotic are employed to control the inflammatory reaction.

Open Globe Injury

Open globe injury is the most devastating form of ocular trauma caused by sharp objects. Even blunt trauma can cause an open globe injury in the eye weakened by previous surgery or trauma. When the impact of blunt trauma is severe, the indirect rebounding compression wave force causes rupture of globe at superonasal limbus, the weakest part of the eye. Prolapse of the uveal tissue and the vitreous may occur at the site of tear. The reconstruction of the anterior segment should be done on an emergency basis.

Extensive open globe injuries, more often than not, cause disorganization of the anterior segment of the eye. Uveocorneal adhesions, occlusio pupillae, more or less total posterior synechia and associated secondary glaucoma cannot be managed by routine surgical procedures. Reconstruction of the anterior segment can be done by pars plana lensectomy, anterior vitrectomy, reformation of the pupil and abscision of the anterior synechia. If the eye is damaged beyond repair, it should be excised.

Penetrating Wounds with Retention of Foreign Bodies

A foreign body may penetrate the eye through its outer coats, the cornea or the sclera, especially when it strikes the eye with a high velocity. The accident commonly occurs in industries. The minute chips of iron, steel, stone, lead pellets, copper, aluminium, glass and wood may be found in the eye. Inert particles (glass, plastic, porcelain) do not induce much reaction, while iron, copper and brass pieces cause both mechanical trauma and chemical reaction in the eye.

The entry of organic foreign bodies such as wood spicules and stones into the eye introduces infection. The presence of eyelash into the anterior chamber may produce a proliferative reaction characterized by the formation of giant cells.

Entrance of Foreign Body

Foreign body may enter the eye either through the cornea or the sclera. When it pierces the cornea in the center, it does not injure the iris but may pass through the lens, vitreous, retina and choroid or even perforate the posterior sclera. A FB penetrating the cornea at the periphery may pass through the iris and the zonule and lie in the vitreous cavity. If a FB passes through the posterior part of the sclera, it enters the choroid, retina and lies in the vitreous. A missile greater than 2 mm in size often causes destruction of the eye.

Site of Lodgement of Foreign Body

Once a foreign body enters the eye it may be lodged at one of the following sites:

1. *Anterior chamber*: A foreign body can sink to the bottom of the anterior chamber or is retained in the angle of the anterior chamber.
2. *Iris*: A foreign body may be found lodged in the iris stroma.
3. *Posterior chamber*: The posterior chamber is an uncommon site for the lodgement of a foreign body.
4. *Lens*: A foreign body may pierce the iris or pass through the pupil to get lodged in the substance of the lens. It leaves behind a hole in the iris and causes formation of a traumatic cataract.
5. *Vitreous*: A foreign body in the vitreous may remain suspended for some time but finally sinks to the bottom of the vitreous cavity owing to liquefaction of the gel vitreous. Fast moving particles traverse the vitreous as well as the retina, choroid and sclera get lodged in the orbital tissue.
6. *Retina*: A retained foreign body in the retina is usually surrounded by hemorrhages and exudates and eventually gets encapsulated. It may be associated with a widespread pigmentary degeneration of the tissue.

Metallic Foreign Bodies

A metallic foreign body (iron or steel) induces specific tissue reaction depending upon its chemical nature. An electrolytic dissociation of metal by the 'current of rest' in the eye disseminates the metal throughout the ocular tissue, the metal particles combine with cellular proteins and cause cell atrophy.

Iron causes *siderosis* which is characterized by appearance of rusty oval patches on the anterior capsule of the lens (arranged radially in a ring corresponding to the dilated pupil), and greenish and reddish-brown discoloration of the iris. The vision deteriorates due to formation of cataract and pigmentary retinal degeneration.

Pure copper foreign body retained in the eye induces violent suppurative reaction which eventually results in shrinkage of the globe. However, copper alloys (brass) induce a mild reaction, *chalcosis*. The dissociated metal particles are deposited in the ocular tissues—Kayser-Fleischer ring in the periphery of the cornea, sunflower cataract and golden-green plaque in the retina. The dissociated copper particles do not combine with the tissue protein, and, hence, degenerative changes do not appear and vision may remain good.

Inert Foreign Bodies

Gold, platinum, glass, plastic and porcelain are inert materials. Retention of these substances in the eye do not induce any reaction.

Nonmetallic Foreign Bodies

Small metallic foreign bodies are often sterile due to heat generated with their commission but pieces of wood or stone usually get contaminated with bacteria and fungi and cause severe ocular infection.

Diagnosis and Localization of Intraocular FB

History: All cases of retained intraocular foreign body (IOFB) should be subjected to a careful history-taking. The type of job in which the patient was involved at the time of accident may give a clue about the nature of foreign body. When a workman uses a hammer and a chisel, it is the minute chip of iron from the mushroomed end of the chisel that enters the eye.

Examination: A clinical examination should be carried out to find the wound of entry. A sclero-

corneal or corneal wound, a hole in the iris or a tear in the sclera and low IOP are suggestive of a retained intraocular foreign body. Small particles can be visualized on slit-lamp examination, but sometimes they may remain hidden in the lower scleral rim**.** Gonioscopy may help to localize the FB in the angle of the anterior chamber.

The examination of fundus using a binocular indirect ophthalmoscope with scleral indentation may detect the foreign body in the posterior segment of the eye (Fig. 22.16) if media are clear. Signs like vitreous tract and retinal hemorrhage help in the localization. The presence of a retinal perforation indicates the lodgement of a foreign body in the sclera or the orbit.

Magnet: When the eye is exposed to a magnet, the feeling of a distinct pull and pain in the eye indicates that the foreign body is metallic. Occasionally, the magnetic foreign bodies are encapsulated and behave like nonmagnetic ones. The application of a magnet for diagnostic purpose is currently not in use. Similarly, Bermans and Roper-Hall locators have only historical importance.

Radiography: A plain X-ray of the orbit may reveal the presence of a FB (Fig. 22.17). In the past, a number of radiography techniques such as plain X-ray with Water's view, X-rays with limbal ring, Comber's method (contact lens marker) and Sweet's method were used to localize an IOFB. However, computerized tomography has replaced these methods.

Computerized tomography (CT) scan: CT scan is a noninvasive procedure which precisely localizes an IOFB (Fig. 22.18). It can localize multiple foreign bodies as well as identify associated intracranial injuries.

Magnetic resonance imaging (MRI): Wooden IOFBs are very precisely localized by MRI. However, MRI is contraindicated in metallic IOFB as the application of a magnetic field can cause move-

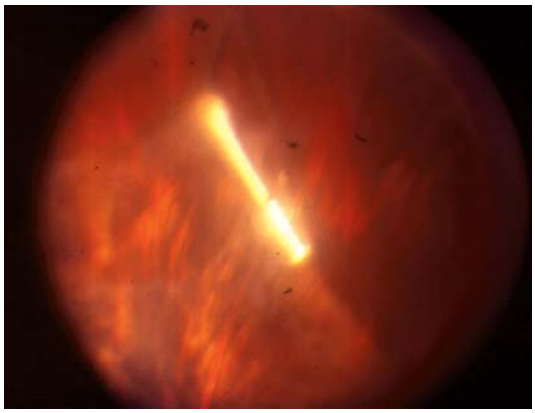

Fig. 22.16: A wire lying on retina (*Courtesy*: Dr Lingam Gopal and Dr Nagpal, Sankara Nethralaya, Chennai)

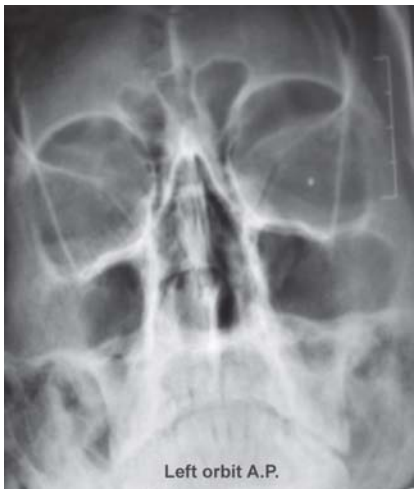

Fig. 22.17: Plain X-ray of orbit showing metallic FB

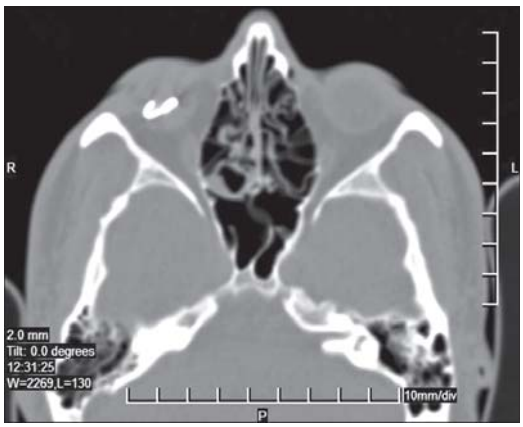

Fig. 22.18: CT scan showing the presence of IOFB (*Courtesy*: Dr MS Bajaj, R P Centre, New Delhi)

ment of the foreign body in the eye and damage the intraocular structures.

Ultrasonography: Ultrasonography can detect both metallic and non-metallic IOFBs. A combination of B and vector A scans can be utilized for localization of a FB (Fig. 22.19). Besides localization of FB, ultrasonography can detect the presence of detachment of the vitreous, choroid and retina, and vitreous incarceration in the posterior tunics of the eyeball at the site of double perforation.

Ultrasound biomicroscopy: IOFB located in the anterior chamber, the iris (Fig. 22.20), the angle of the anterior chamber, and the posterior chamber can be localized with the help of ultrasound biomicroscopy (UBM).

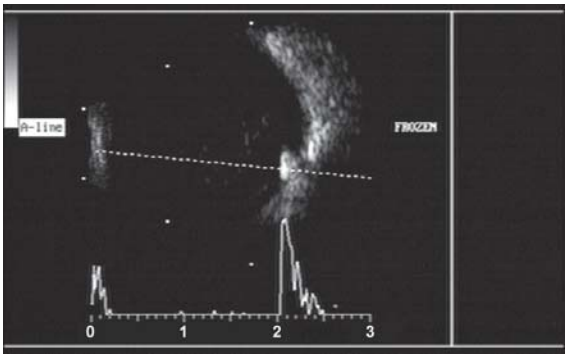

Fig. 22.19: Ultrasonography showing IOFB

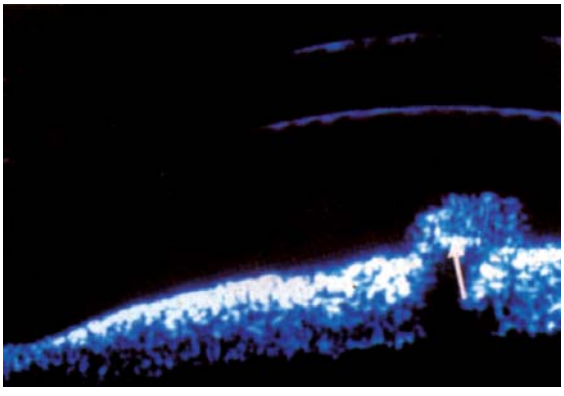

Fig. 22.20: Ultrasound biomicroscopy of FB in iris

Treatment

The removal of an intraocular foreign body is indicated in majority of cases. The procedure is not always simple and may require expertise.

Inert FB

The foreign body may not be extracted if it is an inert one or unlikely to cause damage to the vision. The removal is also deferred when the procedure is likely to destroy the remaining sight in the injured eye.

Magnetic FB

Magnetic foreign bodies lying in the anterior chamber are usually removed under direct vision by employing a magnet after making an incision at the limbus (Fig. 22.21). An iridectomy is needed to remove the foreign body embedded in the iris.

A magnetic foreign body lying within the lens behaves as nonmagnetic and warrants extraction of the opaque lens.

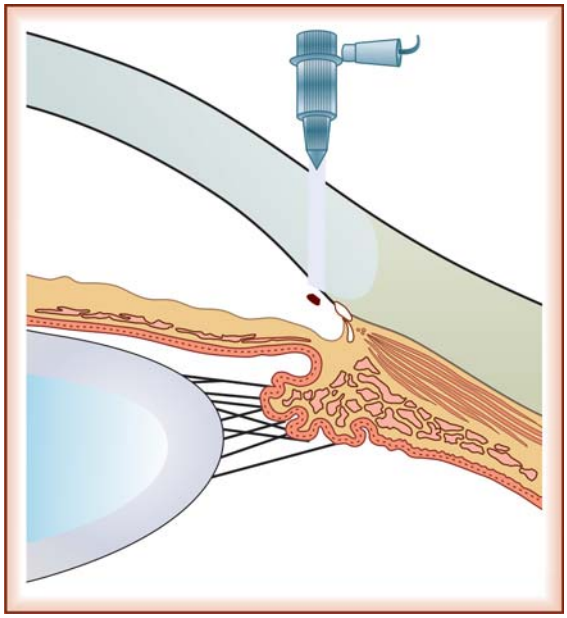

Fig. 22.21: Removal of FB from anterior chamber using a magnet

Magnetic foreign body in the vitreous can be removed either through the anterior route or through the posterior route.

Anterior route extraction: The foreign body is drawn round the lens and into the anterior chamber by a magnet, then it is removed by a forceps from the anterior chamber (Fig. 22.22). This procedure causes extensive damage to the lens and the uveal tissue and is, therefore, suitable for aphakic patients mainly.

Posterior route extraction: A scleral incision is made as close to the foreign body as possible and a magnet is then applied. This approach is useful in the removal of a large jagged foreign body with slight magnetizability (Fig. 22.23).

Nonmagnetic FB

Nonmagnetic foreign bodies from the anterior segment are usually picked out by a forceps after a suitably placed incision. When FB lies in the posterior segment, its removal is accomplished by bimanual vitrectomy using a vitreous cutter and an endoilluminator (light pipe). Intravitreal forceps may be used to pick up small foreign bodies from the vitreous under direct vision using an operating microscope (Fig. 22.24). The extraction of a foreign body from the posterior segment is not without a risk. Many such eyes eventually

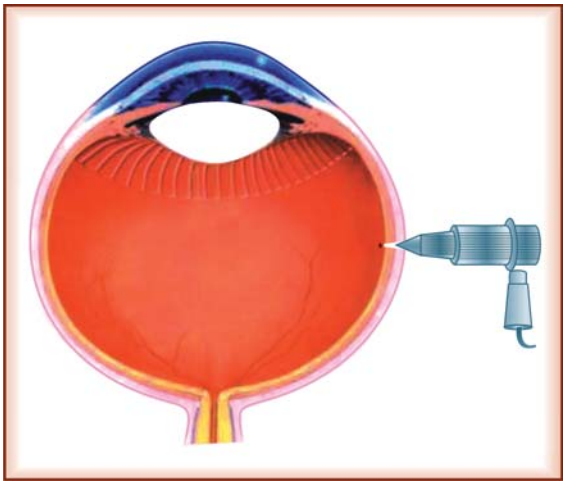

Fig. 22.23: Magnetic posterior segment FB being removed through sclerotomy by a magnet

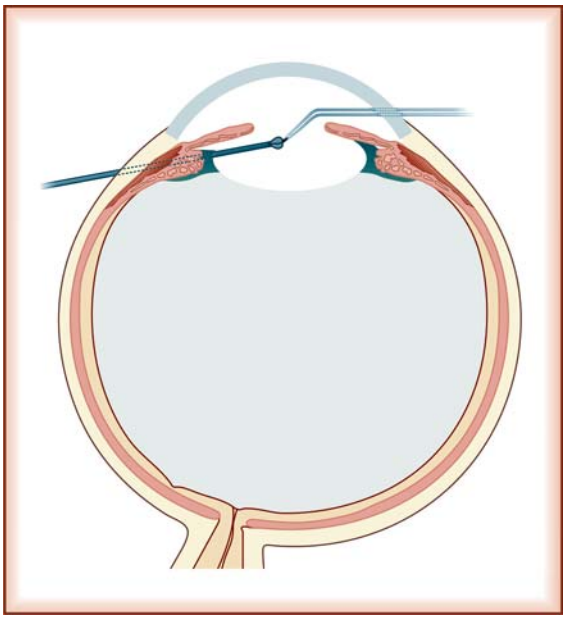

Fig. 22.22: Posterior segment FB being removed from anterior chamber by using a forceps

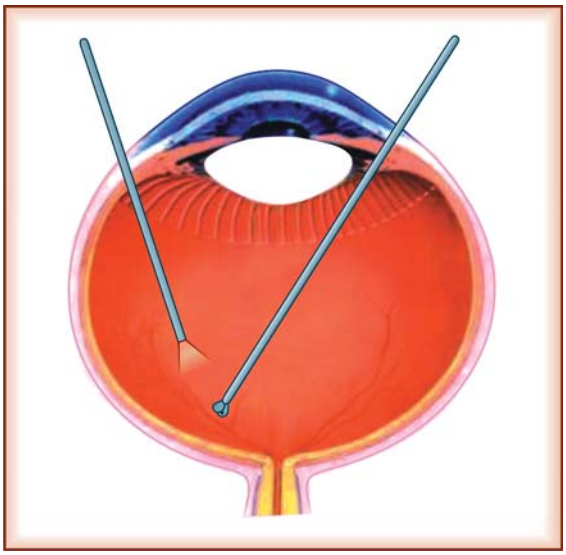

Fig. 22.24: Removal of posterior segment FB by a vitreous forceps

undergo degenerative changes or detachment of the retina and lose useful vision.

Sympathetic Ophthalmitis (Sympathetic Uveitis or Sympathetic Ophthalmia)

Sympathetic ophthalmitis (SO) is a bilateral granulomatous panuveitis that usually occurs in a sound eye (*sympathizing eye)* following perforating injury in the other eye (*exciting eye*). The incidence of the disease has significantly decreased due to improved technique of wound repair and prompt use of corticosteroids and antibiotics.

Risk Factors

1. SO may occur at any age but children are more susceptible.
2. Development of SO is relatively more common following perforating injuries of the ciliary region (danger zone) with incarceration of the uveal tissue and the vitreous.
3. The incidence of SO is high in an eye with retained FB.

Onset

Although the uveitis may start as early as 5 days or as late as 50 years following an injury, majority of patients (80%) develop SO within 3 weeks to 3 months after the ocular trauma. The occurrence of SO is rare if suppuration supervens in the injured eye.

Etiology

The etiology of sympathetic uveitis is yet unknown, it is presumed to be an autoimmune disease. The concept that injury to the uveal tissue could lead to a reaction in the noninjured eye owing to a hypersensitivity to melanin and melanin associated protein is gaining credence. Sensitivity to retinal S-antigen, a soluble protein found in rod outer segments, has been demonstrated in some patients.

Pathology

Histopathology of the exciting as well as the sympathizing eyes shows a diffuse granulomatous inflammation marked by infiltration of lymphocytes, epithelioid cells, eosinophils and a few plasma cells. The epithelioid cells often contain engulfed uveal pigments. The collections of epithelioid cells between the retinal pigment epithelium and the membrane of Bruch, known as *Dalen-Fuchs nodules*, are usually found. The overlying retina and underlying choriocapillaris are spared.

Clinical Features

The onset of sympathetic uveitis is heralded by photophobia, lacrimation and blurred vision in the sympathizing eye and worsening of vision and photophobia in the exciting eye. More often than not, it manifests as granulomatous iridocyclitis that is marked by the presence of ciliary injection, nodular infiltration in the iris, peripheral anterior synechiae, mutton-fat keratic precipitates, tenderness of the eyeball, vitreous opacities and optic disk edema. A fully developed case presents as severe uveitis with plastic exudates in the pupillary region with heavy posterior synechiae and secondary glaucoma. The disease runs a long protracted course but is self-limiting. The exciting eye remains irritable, red and tender with uveitis.

Prophylaxis

Prophylactically, the injured eye should be repaired after excision of the incarcerated tissue and a course of antibiotics and corticosteroids should be administered. Once the injured eye is damaged extensively and cannot regain any vision, it should be excised. The excision of the injured eye, unless sympathetic ophthalmitis has already set in, is a strong safeguard against the disease. When the exciting eye has some useful

vision and sympathetic ophthalmitis has supervened, excision should not be performed because the injured eye may retain better vision than the sympathizing eye.

Treatment

The treatment of sympathetic ophthalmitis is most unsatisfactory.

The essential treatment includes: (i) topical atropine sulphate (1%) drops 4 times a day, (ii) frequent instillations of topical prednisolone acetate (1%) eye drop combined with systemic and periocular administrations of corticosteroids, and (iii) when systemic corticosteroids fail, treatment with antimetabolites is indicated. Cyclosporine is found to be effective in selected patients.

Multiple routes corticosteroids therapy and antimetabolite treatment have improved the prognosis of SO.

CHEMICAL INJURIES

Chemical injuries to eye can occur in home by cosmetics, detergents, disinfectants, solvents, adhesives and drain cleaners. Farmers can be affected by fertilizers and pesticides but serious chemical injuries to the eye result from strong alkalis or acids often in industrial set-up. Alkali burns occur more frequently than acid burns.The incidence of sulfuric acid burn is rising due to the use of battery in inverters.

Alkali Burns

Alkalis (lime, caustic soda, ammonia) may cause dangerous ocular injuries. Initially the burn caused by an alkali may appear less severe but later it leads to marked visual impairment or blindness.

Mode of Action

Strong alkalis cause saponification of fatty acids in the cell membrane and cellular disruption. The damaged surface epithelium allows the alkali to penetrate the corneal stroma and destroy the proteoglycans ground substance and collagen fibers (liquefactive necrosis). The alkali also damages the corneal epithelial stem cells and may penetrate intraocularly.

Acid Burns

Generally acid burns are less dangerous than alkali because most acids do not penetrate deeply into the ocular tissue.

Mode of Action

Acids denature and precipitate the tissue proteins causing immediate coagulative necrosis in the superficial ocular tissue. The precipitated protein acts as a barrier and prevents the acid from penetrating further deep and protecting the proteoglycans ground substance in the cornea from getting damaged. However, acids can incite a severe inflammation that can damage the corneal matrix.

Clinical Features

Profuse watering, blepharospasm, severe pain and visual loss are common ocular symptoms. Acid burn causes immediate visual loss but in alkali burns the loss may occur several days later.

Depending on the severity and duration of exposure to a chemical agent, ocular signs may vary greatly in chemical burns.

1. *Slight degree of burn* causes chemosis of the conjunctiva without any damage to the cornea and the limbus.
2. *Moderate degree of burn* causes damage to the epithelium and the stroma of the cornea and the conjunctiva associated with total corneal erosion, segmental limbal ischemia and mild irritative reaction in the anterior chamber.
3. *Severe degree of burn* causes extensive damage to the conjunctiva, the cornea (Fig. 22.25), the limbal stem cells and the anterior segment of

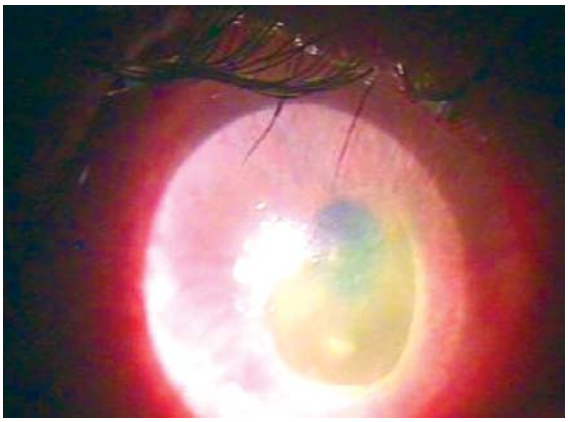

Fig. 22.25: Severe degree of alkali burn

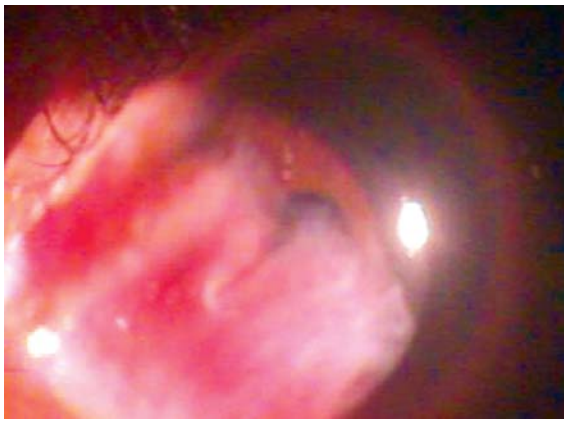

Fig. 22.26: Symblepharon formation following acid burn

the eye. Damage to limbal stem cells results in conjunctivalization of the cornea associated with persistent epithelial defects and vascularization. The eye appears as a *"cooked fish"*. The damage to the conjunctiva results in severe dry eye and symblepharon formation (Figs 22.26 and 22.27). Intraocular penetration of the chemical agent often leads to cataract and secondary glaucoma.

Treatment

The management of chemical injuries include both medical and surgical measures.

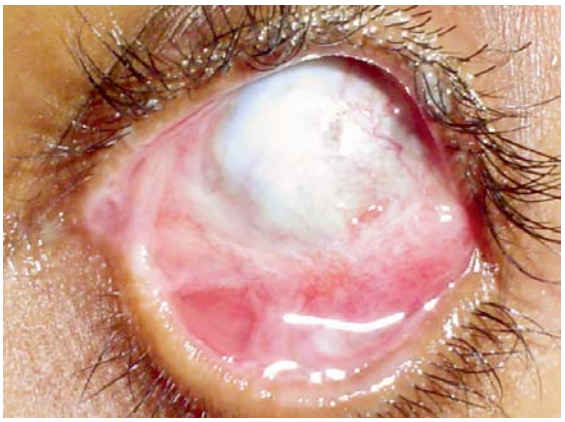

Fig. 22.27: Extensive development of symblepharon following acid burn (*Courtesy:* Prof. Manoj Shukla and Dr Prashant Shukla, AMUIO, Aligarh

Medical

1. Immediate and copious irrigation of eye by normal saline or balance salt solution (BSS) is the most important step in the management of chemical injuries.
2. Removal of particulate chemical from the ocular surface with a swab stick and a forceps after double eversion of the upper eyelid must be carried out.
3. Intensive topical administration of corticosteroids in the acute phase decreases the enzymatic damage to the corneal stroma.
4. Oral tetracyclines are beneficial in inhibiting polymorphonuclear-induced collagenolysis.
5. Topical cycloplegics are indicated in patients with anterior uveitis.
6. High dose of ascorbic acid (1 g per day) promotes collagen synthesis and wound healing.
7. Frequent instillations of nonpreserved lubricants promote epithelial healing.
8. Oral carbonic anhydrase inhibitor controls secondary glaucoma.

Surgical

1. Debridement of the necrotic epithelium allows its replacement by the healthy epithelium.
2. Tarsorrhaphy protects the ocular surface epithelium.
3. Autologous conjunctival transplantion and stem cell implantion from the healthy contralateral eye may restore the corneal epithelium.
4. Amniotic membrane transplantation restores the ocular surface integrity and prevents symblepharon formation.
5. Corneal transplantation may be performed later but has a poor prognosis.

Prognosis

Roper-Halls classification of chemical injuries, based on the corneal appearance and the limbal ischemia, provides prognostic guidelines. Prognosis is good in patients with minimum limbal ischemia while in those with an opaque cornea and ischemia of more than one-half of the limbus have a poor prognosis. Such eyes often end in phthisis bulbi.

Lacrimatory Agents

Lacrimatory gases are aerosol dispersed chemicals that produce ocular and respiratory tract irritation. They are used as riot control agents because of their irritating and incapacitating actions. Common agents used in *tear gas* are chloroacetophenone, chlorobenzylidene nalononitrile or dibenzoxazepine. Exposure to these chemicals causes ocular stinging, pain, excessive lacrimation and inability to open the eyelids. Prompt and thorough rinsing of eye provides immense relief.

Pepper spray or Oleoresin capsicum spray is a lacrimatory agent used for riot control or self defense. It causes irritation to eyes, redness, blepharospasm, pain and profuse watering. The victims should be advised to blink vigorously to encourage tearing in order to flush the irritant from the eye. Irrigation with fresh water, although recommended, has little effect as capsicin is not soluble in water. Flushing the eye with a solution of half *liquid antacid* (of aluminum or magnesium hydroxide base) and half *water* (LAW) is found to be more effective.

Mustard gas (dichlorodiethyl sulphide) is a *poison gas* used as a chemical warfare agent. It causes irritation of conjunctiva and sore sticky eyes on exposure or a chronic and delayed mustard gas keratitis manifesting as recurrent corneal erosions. Prompt irrigation of eyes with running water or normal saline is beneficial. Topical antibiotics, cycloplegic and lubricant are used for the management of corneal erosions.

INJURIES DUE TO PHYSICAL AGENTS

Thermal burns

Etiology

Thermal burns may be caused by cigarette lighters, boiling water, hot ashes, molten metals and gun powder.

Clinical Features

Eyelids are often involved in thermal burns because of the eye-closing reflex. Ectropion of the upper and the lower lid (Fig. 22.28) can occur following burns of the face and the neck.The heat induces coagulation of the corneal and the conjunctival surface. Watering, blepharospasm, visual impairment and pain are the primary symptoms. Particles of ash, metal or gun powder may be found embedded in the cornea, and subsequently the cornea becomes opaque.

Treatment

Superficial particles in the cornea and the conjunctiva should be removed under local anesthesia. Application of cycloplegic agent relieves pain and prevents iridocyclitis. Local and systemic antibiotics prevent infection.

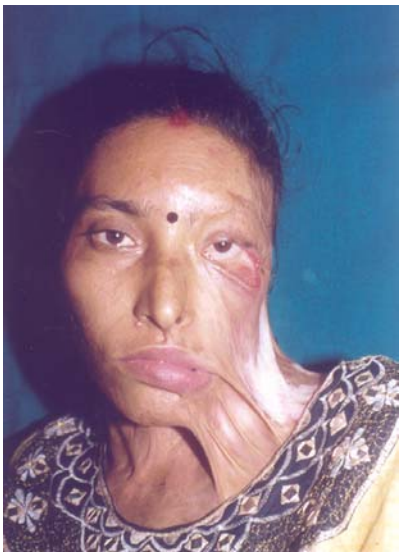

Fig. 22.28: Marked ectropion of lower lid following burn of face and neck (*Courtesy*: Prof. V Bhattacharya, IMS, BHU, Varanasi)

Electrical Injuries

A strong electrical shock can damage the eye. The ocular damage may include corneal opacities, uveitis, cataract, retinal hemorrhages and optic neuritis.

Radiation Injuries

Both ultraviolet and infrared radiations can cause injuries to the eye. The ionizing radiation results in characteristic types of tissue damage which manifests after a latency period.

1. *Ultraviolet radiation*: Exposure to an arc welding and direct or reflected sunlight from snow, especially in mountain climbers, cause ocular discomfort and pain after an interval of a few hours of exposure (snow-blindness). The shedding of corneal epithelium results in superficial ulceration. Use of protective glasses prevents damage. Treatment consists of topical cycloplegic, antibiotic ointment and patching. Some patients may need systemic analgesics as well.
2. *Infrared radiation:* Infrared rays of sunlight are absorbed by the ocular pigment epithelium and cause thermal burns especially photoretinitis *(solar retinopathy)*.
3. *Ionizing radiation:* Ionizing radiation injuries to the eye are observed in patient receiving radiation for the treatment of neoplasm such as tumors of nasopharynx. The radiation causes either a direct tissue damage or a damage to the blood vessels resulting in ischemic necrosis. The ocular features of injury include loss of eyelashes, blepharitis, dry eye syndrome, keratitis, necrosis of the conjunctiva and the cornea, radiation cataract and radiation retinopathy (microaneurysms, flame-shaped retinal hemorrhages, hard exudates, cotton-wool spots, macular edema, arteriolar occlusion and proliferative retinopathy).

   The prophylaxis includes protection of eye before exposure to radiation. Use of bandage contact lens, tarsorrhaphy and frequent instillations of tear substitutes are helpful. Laser photocoagulation for radiation retinopathy and cataract extraction for radiation cataract are recommended.

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CHAPTER

23 Disorders of Ocular Motility: Strabismus

Normally the visual axes of the two eyes remain essentially parallel in all directions of gaze except during convergence. The movements of the two eyes are controlled by voluntary as well as certain reflex mechanisms which, in turn, are governed by centers situated in the brain.

ANATOMY OF EXTRAOCULAR MUSCLES

A set of six extraocular muscles (Fig. 23.1) participate in the movements of each eye. The set consists of four rectus muscles, superior rectus, inferior rectus, medial rectus and lateral rectus, and two obliques, superior and inferior obliques.

Rectus Muscles

The four rectus muscles have a common origin from the annulus of Zinn which encloses the optic foramen.

The superior and medial rectus muscles are closely attached to the dural sheath of the optic nerve which is responsible for the characteristic pain during elevation and adduction in patients with retrobulbar neuritis.

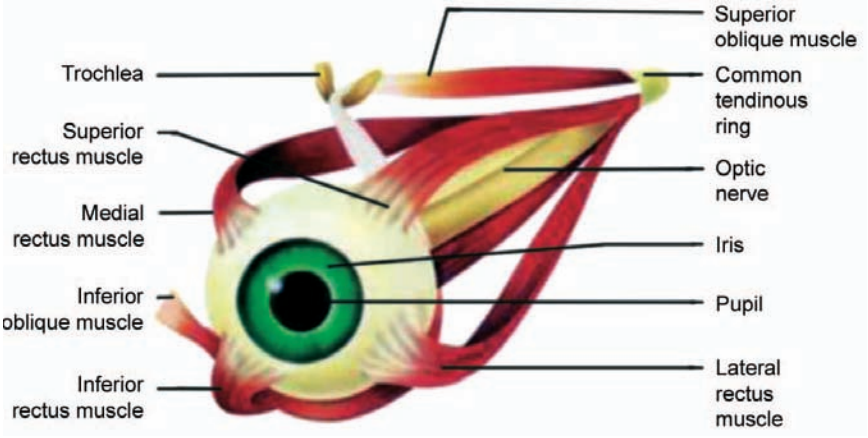


Fig. 23.1: Extraocular muscles (*Courtesy*: Allergan India)

All the four rectus muscles run forwards close to the wall of the orbit and are inserted into the sclera anterior to the equator of the globe by flat tendinous attachments, the average width of the attachments being 10 mm. The medial rectus is inserted into the sclera about 5.5 mm to the medial side of the limbus, the inferior rectus 6.6 mm below, the lateral rectus 7.0 mm to the temporal side, and the superior rectus 7.7 mm above the limbus (Fig. 23.2A).

Oblique Muscles

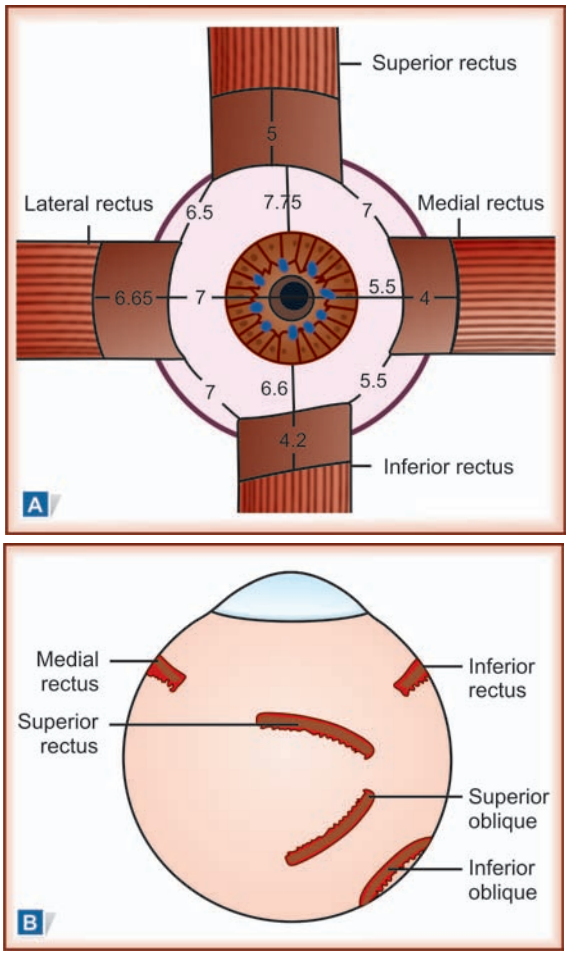
The superior oblique takes its origin from the periosteum of the body of the sphenoid just above and medial to the optic foramen. It runs forwards to the trochlea to pass through it and after becoming tendinous changes its course completely. It runs over the globe posterolaterally underneath the superior rectus and is inserted obliquely in the posterosuperior quadrant of the globe almost laterally.

The inferior oblique is the only extrinsic muscle of the eye which does not take origin from the annulus of Zinn. It arises by a short rounded tendon from a depression on the orbital plate of the maxilla just lateral to the lacrimal fossa. The tendon runs backwards and laterally, passing between the inferior rectus and the floor of the orbit, and is inserted into the sclera in the lower part of posterolateral quadrant of the eyeball.

The line of insertion is oblique with its convexity upwards. The nasal end of the insertion lies just 1 to 2 mm away from the macula (Fig. 23.2B).

Nerve Supply

The third cranial nerve supplies the superior, inferior and medial rectus muscles together with the inferior oblique muscle. The fourth cranial nerve innervates the superior oblique and the lateral rectus is supplied by the sixth cranial nerve.



Figs 23.2A and B: Diagrammatic representation of insertion of extraocular muscles: (A) Insertions of rectus muscles, (B) Insertions of oblique muscles

Blood Supply

The extraocular muscles receive their blood supply from the muscular branches of the ophthalmic artery.

OCULAR MOVEMENTS

The extraocular muscles move the eye around a presumed center situated about 13 mm behind the cornea. Movements of the eyeball take place along three axes.

1. Movements along the *vertical axis* result in pure adduction and abduction of the eyeball.

2. Movements along the *horizontal axis* result in elevation and depression of the eyeball.
3. Movements along the *anteroposterior axis* lead to torsion of the globe.

Terminology of Ocular Movements

When movements are considered in relation to one eye only, they are called *ductions* whereas binocular movements are known as *versions*.

Duction could be inward (adduction), outward (abduction), upward (supraduction) and downward (infraduction).

Version could be conjugate (both eyes moving in unison) or disjunctive (the movements of the two eyes in opposite directions).

The disjunctive movements occur with the axes of two eyes inclined towards each other during convergence and away from each other during divergence.

Positions of Gaze

There are 9 positions of gaze. The following terms are used to describe positions of gaze.

1. *Primary position* is straight ahead.
2. *Secondary positions* are 4 positions of gaze: straight up, straight down, right gaze and left gaze.
3. *Tertiary positions* are 4 oblique positions: up and right, up and left, down and right and down and left.
4. *Cardinal positions of gaze* are 6 in number. These include all gaze positions excluding primary straight, straight up and straight down positions.

Conjugate movements are termed according to the direction of gaze (Fig. 23.3). The terms *dextroversion* and *levoversion* are used for describing the movements of the eyes to the right and left respectively, and the terms *supraversion* and

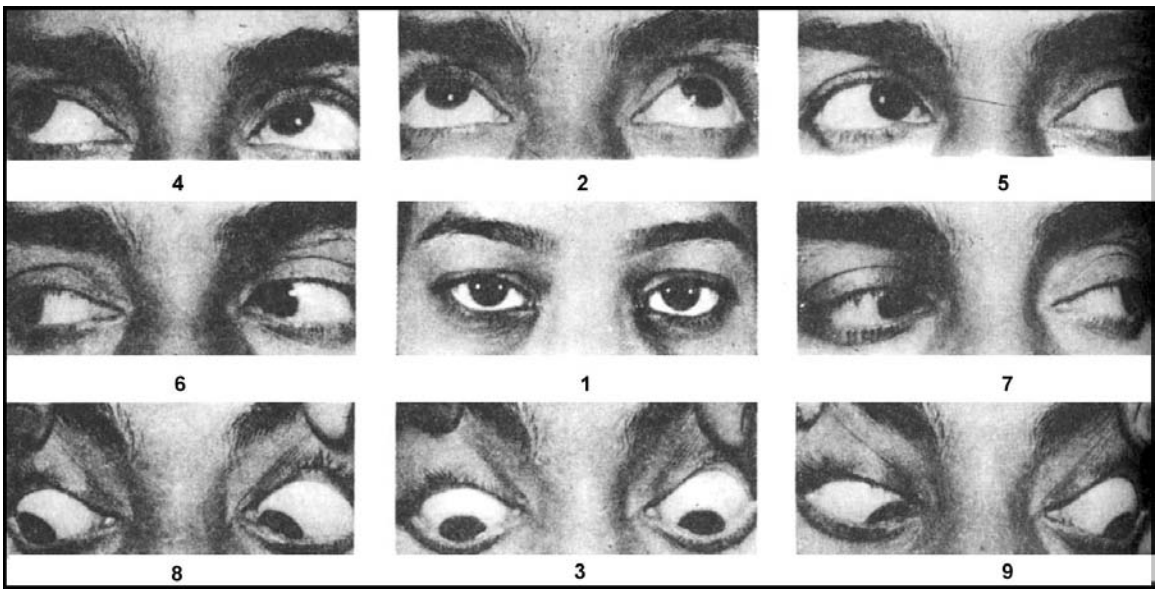


Fig. 23.3: Ocular movements in different gazes. 1. Primary position, 2. Direct elevation, 3. Direct depression, 4. Dextroelevation, 5. Levoelevation, 6. Dextroversion, 7. Levoversion, 8. Dextrodepression, 9. Levodepression

infraversion are for the upward and downward movements, respectively. The torsional movements of both eyes to the right (clockwise) and to the left (anticlockwise) are called *dextrocycloversion* and *levocycloversion*, respectively.

Actions of Extraocular Muscles

The medial rectus is a pure adductor and the lateral rectus abductor. The primary actions of the superior and the inferior rectus muscles are elevation and depression, respectively. The superior rectus has also got subsidiary actions of adduction and intorsion. Similarly, the inferior rectus has got subsidiary actions of adduction and extorsion.

Because of the anatomical course of the muscles, the vertical rectus muscles are elevators or depressors maximally in an abducted position of the eyeball (about 23°).

Since the superior and inferior obliques are inserted behind the globe, their main actions are intorsion and extorsion, respectively. The superior oblique also pulls the eye downwards and the inferior oblique upwards, the maximal action being in the adducted position of the eyeball (about 51°). Both the obliques also aid in abduction (Table 23.1).

Table 23.1: Actions of extraocular muscles

Muscle	*Primary action*	*Secondary action*	*Tertiary action*
Medial rectus	Adduction	–	–
Lateral rectus	Abduction	–	–
Superior rectus	Elevation	Intorsion	Adduction
Inferior rectus	Depression	Extorsion	Adduction
Superior oblique	Intorsion	Depression	Abduction
Inferior oblique	Extorsion	Elevation	Abduction

Agonist, Synergist and Antagonist

A muscle moving the eye in the direction of its action is known as *agonist*. Any muscle which aids the action of some other muscle is called a *synergist* which could either be in the same eye (ipsilateral) or in the other eye (contralateral). Similarly, any muscle opposing the action of other muscle is termed as *antagonist*.

Each extraocular muscle with the exception of medial and lateral rectus muscles has two synergists and two antagonists, while the horizontal rectus muscles have two synergists and three antagonists (Table 23.2).

Yoke Muscles

When the eyes are moving in any of the cardinal positions of gaze, an extraocular muscle of one eye is paired with an extraocular muscle of the contralateral eye. For example, in dextroelevation the two yoke muscles are the right superior rectus and the left inferior oblique and in levoversion the yoke muscles are the left lateral rectus and the right medial rectus. The yoke muscle pairs for six cardinal positions of gaze are listed in Table 23.3.

Table 23.2: Synergist and antagonist of extraocular muscles

Muscle	*Synergists*	*Antagonists*
Medial rectus	Superior rectus Inferior rectus	Lateral rectus Superior oblique Inferior oblique
Lateral rectus	Superior oblique Inferior oblique	Medial rectus Superior rectus Inferior rectus
Superior rectus	Inferior oblique Medial rectus	Inferior rectus Superior oblique
Inferior rectus	Superior oblique Medial rectus	Superior rectus Inferior oblique
Superior oblique	Inferior rectus Lateral rectus	Inferior oblique Superior rectus
Inferior oblique	Superior rectus Lateral rectus	Superior oblique Inferior rectus

Table 23.3: Yoke muscle pairs

Cardinal direction of gaze	*Yoke muscle pairs*
Dextroversion	Right lateral rectus and left medial rectus
Levoversion	Left lateral rectus and right medial rectus
Dextroelevation	Right superior rectus and left inferior oblique
Levoelevation	Left superior rectus and right inferior oblique
Dextrodepression	Right inferior rectus and left superior oblique
Levodepression	Left inferior rectus and right superior oblique

BINOCULAR MOVEMENTS

The binocular movements are governed by two important laws.

1. Hering's law of simultaneous and equal innervation, and
2. Sherrington's law of reciprocal innervation.

Hering's law states that during conjugate ocular movement, simultaneous and equal innervation flows to the yoke muscles.

Sherrington's law states that increased innervation and contraction of an extraocular muscle are accompanied by a reciprocal decreased innervation and contraction of its antagonist.

Both the above laws simplify to a large extent the complexities of ocular movements and explain certain changes occurring in muscles in incomitant strabismus or long-standing cases of comitant strabismus. For example, when the eyes are turned to the right (dextroversion), the right lateral rectus and left medial rectus muscles contract due to an equal and simultaneous flow of innervation and the right medial rectus and left lateral rectus muscles receive inhibitory impulses that cause them to relax.

Hering's law has significant clinical application in the diagnosis of the paralytic strabismus. The amount of innervation flowing to both the eyes is always determined by the fixing eye. For example, in a case of right lateral rectus palsy there is an inward deviation of the right eye owing to unopposed action of the right medial rectus (antagonist). During dextroversion, normal innervation (+) is needed to move the left eye in adduction, but the right eye does not move beyond the midline since the normal amount of innervation (+) cannot overcome the paresis of right lateral rectus. In cover-un cover test, when the sound eye fixates, the deviation shown by the paralyzed eye is called the *primary deviation*. When the paretic right eye is forced to fixate, excessive innervation (+++) is required to abduct the eye. According to Hering's law the same amount of innervation (+++) flows to the normal medial rectus of the left eye resulting in excessive adduction. If the cover is removed, the left eye moves laterally to take up the fixation. This deviation of the sound eye is known as *secondary deviation*, which is always greater than the primary in the paralytic strabismus.

Sherrington's law of reciprocal innervation states that during ocular movements, increased contraction of an extraocular muscle is accompanied by a decreased contractile activity of its antagonist. For example, in dextroversion the contraction (+) of right lateral rectus and left medial rectus is accompanied by decreased tonus (0) of their antagonists—right medial rectus and left lateral rectus. Exceptions to Sherrington's law do occur in physiological conditions like during convergence and divergence.

BINOCULAR VISION

Binocular vision is defined as the vision achieved by the co-ordinated use of both eyes in such a way that the images formed in individual eyes separately are appreciated as a single mental picture. It is gradually acquired and reinforced during the first few years of life. The development

of binocular vision is dependent on the following three factors:

1. Adequate degree of central and peripheral visions in both eyes
2. Perfect motor mechanism to maintain the two eyes in a correct positional relationship at rest and during movement, and
3. A central (cortical) mechanism to promote fusion of two slightly dissimilar images.

Normally, the two visual axes are involuntarily so adjusted that the sharp image of an object is formed on the macula of each eye (*simultaneous perception*). Other adjoining objects form retinal images upon the temporal side of the retina of one eye and upon the nasal side of the other. These retinal areas are co-ordinated in the visual cortex so that a single image of the object is perceived. This is known as *fusion*.

The visually co-ordinated points on the retina are called *corresponding points*. The most important pair is the foveae. Noncorresponding points on the retina are called *disparate points*, the retinal images formed on disparate points may not be fused and are thus seen double (diplopia). If disparity is minor, there is a tendency to fuse the images by means of cortical fusional reflexes. As the most sharp vision is attained by the foveae, the eyes are so oriented that the image of an object falls upon them. This orientation is called *fixation reflex* and can be demonstrated by induced optokinetic nystagmus. The optokinetic nystagmus can be utilized in testing the integrity of the vestibulo-ocular reflex pathway and visual acuity in infants.

Fig. 23.4: Synoptophore

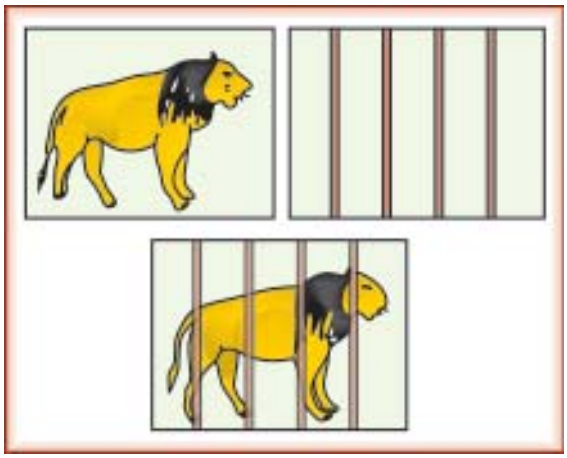


Fig. 23.5: Simultaneous macular perception slides

Grades of Binocular Vision

Binocular vision is precisely measured on a synoptophore (Fig. 23.4). It is of three grades.

Grade I: Simultaneous Macular Perception

When dissimilar test targets, a lion on one side and a cage on the other (Fig. 23.5), are presented the subject sees the lion as being in the cage. If one object is not seen, the eye on the corresponding side is suppressed.

Grade II: Fusion

Two similar targets, which individually lack some detail but when superimposed form a complete

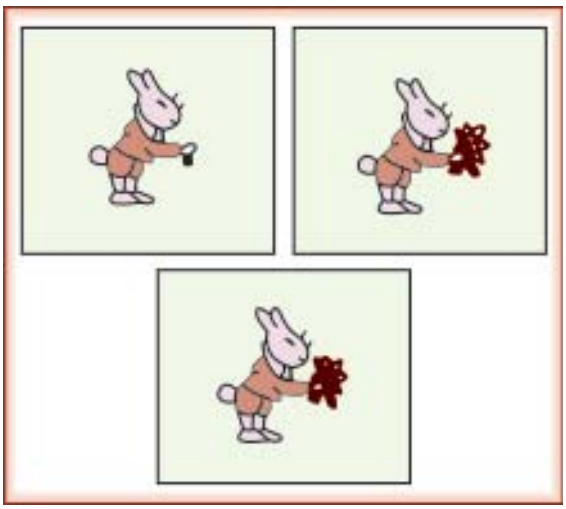


Fig. 23.6: Fusion slides

image, are used for the assessment of fusion (Fig. 23.6). If a complete image is perceived and maintained despite the targets being moved on either side from 5 to 10 degrees, fusion is present.

Grade III: Stereopsis

The third grade of binocular vision is stereopsis or three-dimensional vision. It is the ability to obtain an impression of the depth by the superimposition of two images of the same object taken from slightly different angles. The stereopsis can be measured by stereopsis slides (Fig. 23.7).

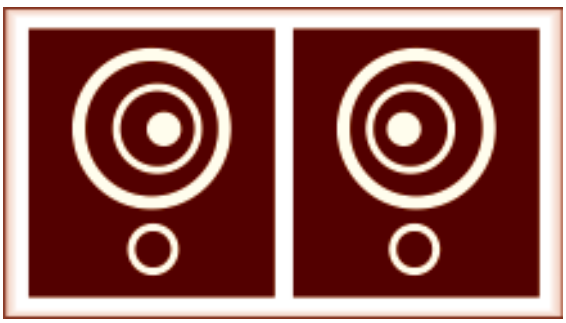


Fig. 23.7: Slides to elicit stereopsis

Development of Binocular Vision

The development of binocular vision is a gradual process. At birth, the movements of the two eyes are not co-ordinated. Conjugate fixational reflexes develop by the fourth week after birth. Binocular fusional reflexes develop by the age of six months. The corrective fusional reflex, which allows the eye to function binocularly even under conditions of stress, starts functioning during the first year of life and by the age of 5 to 6 years is fully established.

Abnormalities of binocular vision include suppression, amblyopia, abnormal retinal correspondence and eccentric fixation.

When one eye deviates, the image of an object falls on an extrafoveal area of the deviating eye resulting in diplopia. This causes confusion and disorientation. To get relief from diplopia, there is an attempt to suppress the image in the deviating eye. This suppression later deepens into amblyopia.

Sometimes, an extrafoveal point of the deviating eye co-ordinates with the fovea of the fixing eye in binocular vision. This is called *anomalous retinal correspondence*. Occasionally, when the normal eye is covered, the deviating eye continues to fix by an extrafoveal area resulting in eccentric fixation.

STRABISMUS (SQUINT)

Normally, the image of an object of regard falls on the fovea of each eye, but certain eyes are so positioned that the image falls upon the fovea of one eye but not on the fovea of the other. This condition where there is misalignment of the visual axes of the two eyes is called *strabismus* or *squint*.

Classification of Strabismus

Strabismus may be classified on the basis of age of onset, type of deviation, fusional status and variation of deviation with gaze position.

1. *Age of onset*: (a) Congenital, and (b) Acquired
2. *Type of deviation:* (a) Horizontal, (b) Vertical, (c) Tortional, and (d) Combined
3. *Fusional status*: (a) Phoria with fusional control, and (b) Tropia without fusional control
4. *Variation of deviation with gaze position*: (a) Comitant, and (b) Incomitant.

Pseudostrabismus

Sometimes, pseudostrabismus may be present due to the presence of certain anomalies, viz., epicanthal folds or abnormal angle kappa. An apparent divergent strabismus is found in high hypermetropia as a result of positive angle kappa, while a negative angle in high myopia gives an apparent convergent strabismus.

The *angle kappa* is defined as the angle between the visual axis (line connecting the point of fixation with the fovea through the nodal point) and the pupillary axis (line passing through the center of the pupil perpendicular to the cornea) (Fig. 23.8). Clinically, the angle is measured on the corneal surface. The angle is positive when the light reflex is found nasally and negative when it is present temporally. An angle kappa of 5° (positive) is usually found in an emmetropic eye.

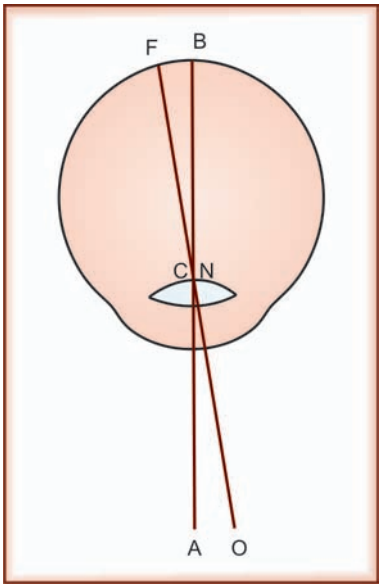


Fig. 23.8: Angle kappa: O: Object, N: Nodal point, F: Fixation point, A-B: Pupillary axis, O-F: Visual axis, ONA: Angle kappa

Incomitant Strabismus (Paralytic Strabismus)

Incomitant strabismus is characterized by impaired action of one or more extraocular muscles associated with diplopia and variation in the angle of deviation in different directions of gaze.

Etiology

Incomitant strabismus may be caused by neurogenic, myogenic or mechanical causes.

Neurogenic Causes

The lesions of nerves supplying the ocular muscles can occur at various levels. Infranuclear lesions of the trochlear and the abducent nerves are common. Nuclear lesions produce ocular muscle palsies. Supranuclear lesions cause conjugate gaze palsies.

1. *Inflammatory lesions:* Multiple sclerosis, encephalitis, poliomyelitis and meningitis may cause paralytic strabismus. Nerve trunk supplying the extraocular muscles may be involved in the infectious lesion of cavernous sinus and orbit. Multiple sclerosis and infectious diseases often implicate the nerve supplying the extraocular muscle in young patients.
2. *Trauma:* Head injury is a common cause of the abducent nerve palsy.
3. *Vascular accidents:* Small hemorrhages and thrombotic lesions of the midbrain may occur in older patients. Such accidents may be associated with hypertension, diabetes and atherosclerosis.
4. *Neoplasm:* Brain tumors and malignant nasopharyngeal growth can produce ocular muscle palsies.

5. *Toxins:* Diphtheria, botulinum toxin and lead poisoning may lead to incomitant strabismus.
6. *Congenital anomalies*: Congenital anomalies of the extraocular muscles and their fascial attachments may lead to incomitant strabismus.

Myogenic Causes

Myogenic causes are not common and these include:

1. Graves disease
2. Myasthenia gravis
3. Chronic progressive ophthalmoplegia
4. Ocular myositis.

Mechanical Causes

Blow-out fracture of orbit, hematoma and swelling in the orbit and facial bones may restrict the extraocular muscle movements causing incomitant strabismus.

Clinical Features

Diplopia and vertigo are the most distressing symptoms of incomitant strabismus.

Diplopia: Diplopia occurs in the field of action of the paralyzed muscle i.e., the direction in which the primary action of the muscle is greatest. The image seen by the squinting eye (false image) is often less distinct than that seen by the sound eye (true image). The diplopia may be homonymous (uncrossed, Fig. 23.9A) or heteronymous (crossed, Fig. 23.9B). Depending on the muscle involved it may be horizontal or vertical.

Vertigo: Vertigo leading to nausea and vomiting is due partly to diplopia and partly to false projection. It is maximal when the patient looks in the direction of the action of paralyzed muscle. However, it is minimized by changing the head posture or completely eliminated by closing the

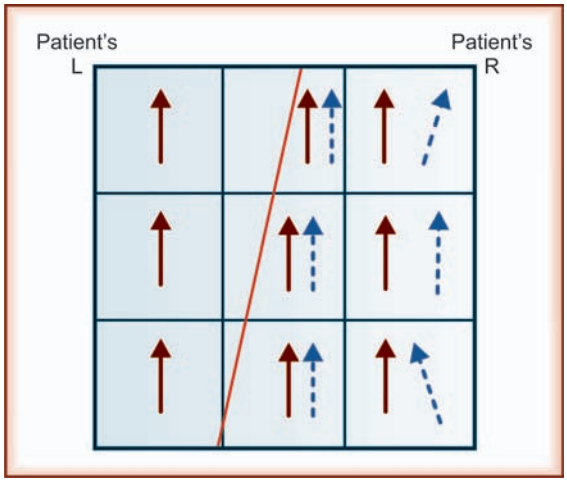


Fig. 23.9A: Homonymous diplopia in right lateral rectus paralysis

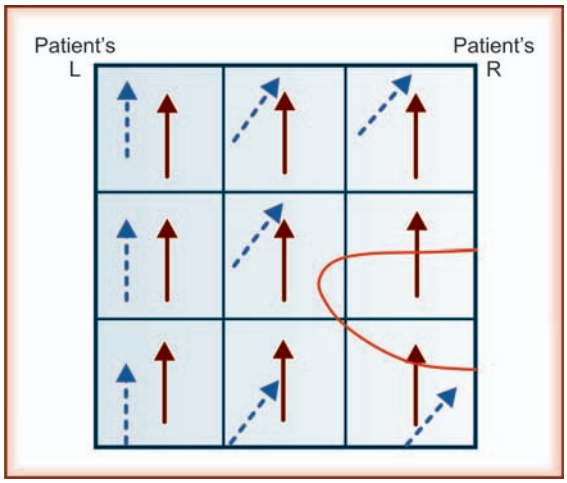
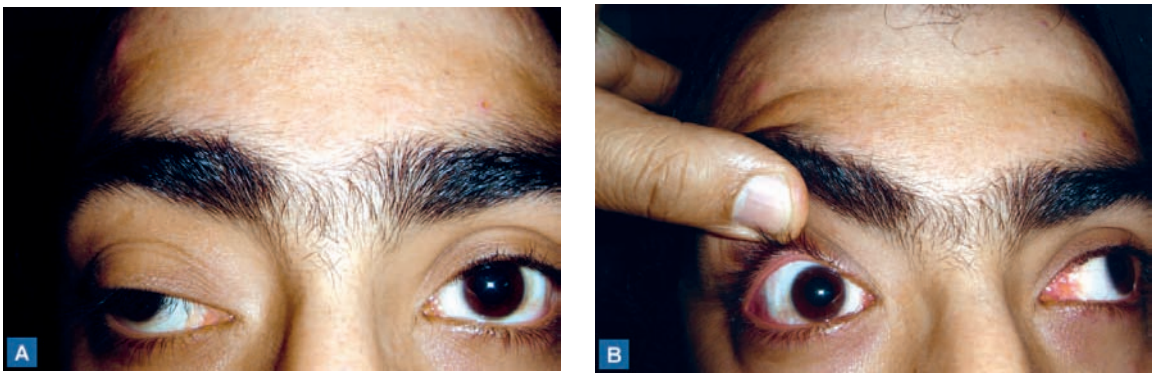


Fig. 23.9B: Heteronymous diplopia in right third nerve paralysis

affected eye. These symptoms are absent or not so alarming in congenital incomitant strabismus as the vision in the affected eye is invariably poor or because of development of anomalous retinal correspondence.

Abnormal deviation of the eye, limitation of ocular movements, abnormal positioning of the head and false orientation are the important signs of the incomitant strabismus.



Figs 23.10A and B: Right third nerve paralysis: (A) Right divergent strabismus, (B) Limitation of action of right medial rectus muscle on levoversion. (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

Angle of deviation: In incomitant strabismus, the affected eye gets deviated, for example, it turns inwards in external rectus paralysis and turns outwards (Fig. 23.10A) in medial rectus paralysis. The angle of deviation is the angle which the line joining the object of regard and nodal point makes with the visual axis. The angle of strabismus varies in different gazes.

Primary and *secondary deviations*: When the sound eye fixates, the deviation shown by the squinting eye is called *primary deviation*. On the other hand, if the paralyzed eye is forced to fixate by covering the sound eye by an occluder, the deviation shown by the sound eye, when uncovered, is called *secondary deviation*. The secondary deviation is always greater than the primary deviation in incomitant strabismus. This can be well explained on the basis of Hering's law.

Limitation of ocular movements: Ocular movements are often restricted in the direction of action of the paralyzed muscle (Fig. 23.10B).

Compensatory head and *chin position:* In incomitant strabismus, the patient adopts a compensatory head posture. For example, in paralysis of right lateral rectus, the patient keeps his head turned to the right (in the direction of action of the paralyzed muscle) as a compensatory maneuver to avoid diplopia. In the paralysis of vertically acting muscles, the head tilting phenomenon is associated with depression or elevation of the chin. For example, in a case of right superior oblique palsy, the head is tilted to the left and the chin depressed, thus the eyeballs are directed up and right (dextroelevation). The right superior oblique being a levodepressor remains in this position in a state of relaxation.

Ocular torticollis: Tilting of the head to compensate for defective vertical movements of the paretic eye is known as *ocular torticollis*. It is found in congenital or traumatic palsies to avoid diplopia. However, the diplopia can be elicited and the vertical strabismus is made manifest by keeping the head straight. The ocular torticollis must be differentiated from true torticollis. In true torticollis, there occurs undue contraction of the sterno-mastoid muscle and the head is tilted with rotation of the chin to the opposite side.

False projection or *false orientation:* It unables the paralyzed eye to locate the objects in space correctly. The false projection depends upon the same principle as the secondary deviation. The objects are usually projected too far in the direction of the action of the paralyzed muscle due to greater flow of innervational stimulus than that required in normal circumstances.

Ophthalmoplegia: In paralytic strabismus one or more extraocular muscles may be affected. Isolated palsy of lateral rectus is quite common. In complete third nerve palsy, besides the involvement of medial rectus, superior rectus, inferior rectus and inferior oblique muscles, levator and sphincter pupillae also occurs. The paralysis of the extrinsic as well as intrinsic muscles of the eye is called *total ophthalmoplegia.* When only the external muscles are affected, the condition is known as *external ophthalmoplegia* and if only the intrinsic, *internal ophthalmoplegia.*

Postparalytic Secondary Changes in Extraocular Muscles

Certain secondary changes always take place in a long-standing ocular muscle palsy (paralytic strabismus) particularly due to the nerve lesion. These changes or sequelae are not confined to the muscle of the affected eye but also involve the contralateral eye. The changes following single extraocular muscle palsy are as follows.

1. Overaction of the contralateral synergist
2. Contracture of the ipsilateral direct antagonist
3. Secondary inhibitional palsy of the contralateral antagonist.

For example, in a case of right lateral rectus palsy following changes ensue:

1. Overaction of left medial rectus
2. Contracture of right medial rectus
3. Secondary inhibitional palsy of left lateral rectus.

Interestingly, if these changes reach a stage of complete balance, they may sometimes acquire the features of comitant strabismus.

Investigations

The cause of incomitant strabismus should be probed by history, systemic examination and imaging techniques. Besides routine investigations, each case of incomitant strabismus should be investigated for the type of diplopia, secondary changes in muscles and entrapment of muscle by the following tests.

1. *Diplopia charting:* It is done in a semi-dark room. The patient is asked to wear red-green glasses and shown a candle light or a slit torchlight from a distance of 4 feet. The position of images are recorded in all the gaze positions.
2. *Hess screen test:* The test is performed to chart the fields of two eyes which may demonstrate overaction, contracture and secondary inhibitional palsy of the involved muscle.
3. *Forced duction test:* The test differentiates between the restriction of ocular movement due to mechanical causes such as entrapment of the muscle in the fractured floor of the orbit and the extraocular muscle palsy.

Treatment

Every case of incomitant strabismus should be managed initially on conservative lines. The treatment is directed to the cause of palsy.

Conservative measures: Troublesome diplopia can be controlled by occluding the paretic eye or by prescribing a prism. No surgical intervention should be done within six months of the paralysis as many cases are known to recover during this period. By this time many paralytic cases may acquire the features of non-paralytic (comitant) strabismus and this change is a safe indication for surgical intervention.

Chemodenervation: Botulinum toxin injection is found to be effective in the following conditions: (i) small to moderate angle esotropia and exotropia, (ii) postoperative residual strabismus, and (iii) acute VI cranial nerve palsy.

Surgery: The main aim of the surgical treatment of incomitant strabismus is to restore a comfortable binocular vision in the primary position. The cases with paresis of medial and lateral rectus

muscles are managed on more or less similar lines as those of comitant strabismus. For example, in a case of paresis of right lateral rectus, weakening of the overacting contralateral synergist (left medial rectus) gives better results than the weakening of ipsilateral direct antagonist (medial rectus of the right eye). However, some cases may need an additional surgery such as recession of right medial rectus or resection of right lateral rectus.

In complete paralysis of a muscle, the muscle transplant operation is indicated. For example, in a case of complete paralysis of lateral rectus muscle, half of the superior rectus and half of the inferior rectus tendons are transplanted to the insertion of the lateral rectus combined with the recession of the medial rectus.

In everyday life we are more concerned with looking downwards than upwards. Therefore, an imbalance of the vertically acting muscles accentuated by downward gaze is very distressing. Utmost care should be exercised while operating upon a depressor muscle. It is preferred to operate upon an elevator muscle rather than on a depressor.

Comitant Strabismus

Incomitant strabismus although the eyes are misaligned they maintain their abnormal relationship in all directions of cardinal gaze. In contrast to the defect in the efferent mechanism found in incomitant strabismus, the efferent pathways in comitant strabismus are normal and thus the eyes retain their coordination.

Etiology

Comitant strabismus frequently occurs in children and manifests within the first two years of the life. The causes of the strabismus are often unclear. However, following factors have been identified.

1. *Genetic:* Family history of strabismus is found in approximately 60% of the patients. In some cases, the strabismus is transmitted as an autosomal dominant trait. Siblings may inherit the same type of ocular deviation as their parents have or had.
2. *Uncorrected refractive error:* It is an important factor responsible for the occurrence of comitant strabismus.
3. *Convergence:* An increased or decreased convergence may be associated with comitant strabismus.
4. *Imbalance between accommodation and convergence:* Forced dissociation between accommodation and convergence may cause strabismus. Children with hypermetropia have to accommodate constantly to see clearly the distant objects. The accommodation tiggers an excessive convergence impulse causing esophoria. On the other hand, high myopes develop exophoria due to lack of accommodative impulse.
5. *Anisometropia*: Anisometropia and anisiekonia cause insufficient fusion resulting in phoria or tropia.
6. *Unilateral visual impairment:* The presence of an opacity (corneal opacity or congenital cataract) in the ocular media forces the child to use one eye only resulting in loss of fixation of the affected eye and its subsequent deviation.
7. *Congenital and developmental defects of extraocular muscles:* The congenital defects may cause muscular imbalance and lead to comitant strabismus.
8. *Defects in the central mechanism*: The central defects controlling the fixation and fusional reflexes may result in comitant strabismus.

Types

Comitant strabismus is usually divided into two broad categories:

1. Heterophoria (Latent strabismus), and
2. Heterotropia (Manifest strabismus).

Heterophoria

Heterophoria (*latent strabismus*) is a condition in which there is a tendency for nonalignment of the visual axes which is corrected or compensated by the fusional reflex. When the two eyes are dissociated by covering one eye, the deviation gets manifest in the covered eye. Maintenance of a perfect alignment of the visual axes in different gazes is a rarity and some degree of heterophoria is universal.

Heterophoria can be of 4 types:
1. Esophoria
2. Exophoria
3. Hyperphoria, and
4. Cyclophoria.

Esophoria

The eyes have a tendency to converge. On the basis of etiology it may be subdivided into two categories:
1. *Convergence excess type:* It causes greater esophoria for near than for distance, and
2. *Divergence weakness type:* It presents greater esophoria for distance than for near.

Usually there is a tendency for esophoria in childhood since developmentally the medial rectus muscle is stronger than the lateral rectus.

Exophoria

The eyes have a tendency to diverge. It is also subdivided into two categories:
1. *Convergence weakness type:* It cause greater exophoria for near than for distance, and
2. *Divergence excess type:* It presents greater exophoria for distance than for near.

Hyperphoria

The eyes have a tendency to deviate vertically resulting in vertical misalignment of the visual axes. The vertical deviations are expressed in terms of the relative position of the two eyes. Conventionally, it is expressed as hyperphoria of one eye.

Cyclophoria

Cyclophoria is a tortional rotation of eyes occurring around the anteroposterior axis of the eyeball. It is also sub-divided into two categories:
1. *Incyclophoria* or *intortion*: The vertical meridian of the cornea rotates nasally, and
2. *Excyclophoria* or *extortion:* The vertical meridian of the cornea rotates temporally.

In all phorias, the deviation is equally shared between the two eyes. This fact must be remembered while treating the condition.

Clinical Features

Eyestrain, headache and pain in the eyes are important symptoms of heterophoria (especially of larger degrees, 10° or more). Small degrees of esophoria or exophoria give little trouble but the slightest degree of hyperphoria causes discomfort. The discomfort is maximal in cases of cyclophoria.

The symptoms of heterophoria are marked, particularly when the demand for near work is called for. Blurring and running of letters, momentary diplopia and headache are common. Some cases of phoria may become manifest during ill-health, fatigue and anxiety; such deviations are known as *periodic strabismus*.

Diagnosis

Heterophoria can simply be diagnosed by abolishing the fusion reflex. Alternate cover test, Maddox rod test and Maddox wing test can be employed for the detection and measurement of heterophoria.

1. *Alternate Cover Test*

The patient is asked to look at a distant object. Usually, there is no deviation. One eye is then covered by an occluder. The occluded eye deviates. When the occluder is removed the deviated eye moves at once to regain the position of binocular fixation. The test is repeated on the other eye which reacts similarly. The alternate cover test must be done for near as well. The fast or slow recovery of the deviated eye is also noted on removal of the cover.

2. *Maddox Rod Test*

The patient is seated 6 meters away from a spot light in a dark room. A Maddox rod (Fig. 23.11A) is placed horizontally before the right eye, the spot light appears as a red vertical line. The other eye is kept open. If the eyes are orthophoric, the vertical red line seen by the right eye passes through the center of the spot light seen by the left eye. When the line is on the left of the spot light, there is exophoria and if on the right, esophoria is diagnosed.

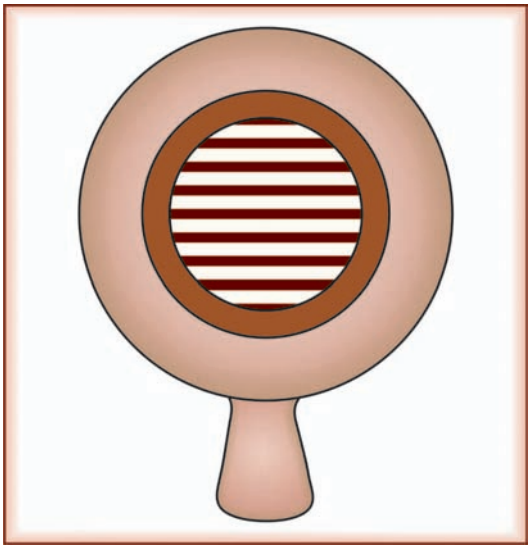


Fig. 23.11A: Maddox rod

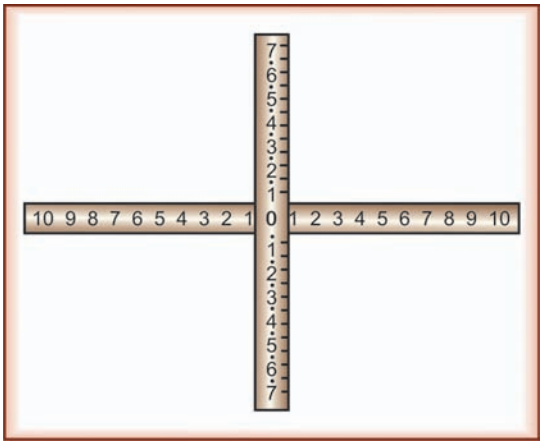


Fig. 23.11B: Tangent scale

Vertical phorias are tested by the rotation of Maddox rod vertically so that the red line becomes horizontal. In vertical deviation, the red line is either below or above the spot.

The degree of phoria can be read directly on a tangent scale (Fig. 23.11B). The phoria can be more correctly measured by placing a suitable prism before one of the eyes so that it brings the vertical line in the center of the spot light.

3. *Maddox Wing Test*

Maddox wing (Fig. 23.12) is used to measure the degree of deviation in prism diopters for near vision. When the patient looks through the eyepieces, the right eye sees a white arrow pointing upwards and a red arrow pointing horizontally to the left. The left eye sees a horizontal scale in white and a vertical scale in red. The fields exposed to each eye are separated by a diaphragm in such a manner that they glide tangentially into each other. Initially, both the arrows should be at zero. The type and degree of deviation can rapidly be determined from the apparent position of the arrow on the scale. A small degree of exophoria (6-8 prism diopters) is of no clinical significance in an adult patient. It simply indicates a diminished accommodative convergence.

4. *Prism Bar Test*

In heterophoria, the strength of the muscles involved can be tested by forcing them to a

Fig. 23.12: Maddox wing

maximal effort against prisms. The patient is seated 6 meters away from a light source, a prism bar is placed before his eye. The strongest prism permitting a single vision provides the vergence power for the particular direction. The test provides a useful information about the amplitude of convergence and divergence in a patient. When convergence falls below 20° (normal convergence 50°), it indicates a convergence deficiency. The normal divergence is 4-5°.

5. *Synoptophore*

The fusional reserve is measured on synoptophore. The normal value of horizontal positive fusional reserve is 20 to 40° while the negative is 3 to 5°. The vertical reserve is only 1.5 to 2.5°.

Treatment

Generally, small degrees of esophoria or exophoria do not warrant any treatment. The refractive error should be properly corrected. In case of asthenopia with convergence deficiency, the amplitude of convergence can be increased by a simple exercise using a pencil.

Pencil push-ups: A pencil is gradually brought towards the nose until its tip is seen double. The patient is advised to look at a distant object after every 30 seconds to relax the accommodation. As a routine, the exercise should be carried out at least three times a day for 3 weeks.

Exercise on synoptophore: The exercises can also be done on a synoptophore for improving the convergence and the fusional reserve.

Prism: When no improvement is obtained by exercises, the asthenopia is often ameliorated by ordering relieving prisms with apex towards the direction of deviation.

Surgery: The large degree of deviation is corrected by surgery.

Heterotropia

Heterotropia (*manifest strabismus*) is a condition wherein abnormal deviation of the visual axes occurs, however, visual axes retain their abnormal relation to each other in all directions of gaze.

Etiology

The etiology of heterotropia is not known. Perhaps, the fusional reflexes are weakly developed or not at all developed resulting in ocular deviation. Anisometropia and opacities of the cornea or lens are some of the conditions which predispose to nonfixation of the involved eye. Muscular imbalance (due to congenital or developmental ocular muscle anomalies) and forced dissociation between accommodation and convergence tend to dissociate the two eyes.

Types

Comitant strabismus may be intermittent (periodical), constant and alternate types.

In a *unilateral strabismus*, one eye habitually deviates and the other fixates. When on alternate cover test, the fixation is retained by either eye, the deviation is known as *alternating strabismus* (Figs 23.13A and B).

On the basis of the direction of deviation, the heterotropia is classified as:

1. Esodeviation (convergent)
2. Exodeviation (divergent)
3. Hypertropia, and
4. Mixed deviation.

Esodeviation

The esodeviation (Fig. 23.14A) is frequent and more common in hypermetropes. It usually starts

Figs 23.13A and B: Alternating convergent strabismus. (A) Right eye deviates inward while left eye fixates, (B) Left eye deviates inward while right eye fixates

in childhood and occasionally manifests after an attack of a debilitating disease. Visual acuity becomes reduced in the deviating eye in uniocular convergent strabismus, while it is not affected in alternators.

Types of Esodeviation

The esodeviation can be divided into following types depending upon its association with refractive error and the amount of accommodative convergence per unit of accommodative response (AC/A ratio):

1. ***Infantile esotropia:*** It has an onset at birth and often associated with nystagmus and neurologic and developmental disorders. The misalignment is larger than 30$^{\Delta}$ (2 prism diopters of deviation is approximately equal to 1 degree). Surgery is needed for the ocular alignment.
2. ***Accommodative esotropia:*** It has an onset around 2 years of age and usually associated with amblyopia. Accommodative esotropia is subdivided into three types:
    a. *Refractive accommodative esotropia (Fig. 23.14B)* results from uncorrected hypermetropia and insufficient fusional divergence. The uncorrected hypermetropia forces the patient to accommodate, thus leading to an increased convergence. The amount of hypermetropia is about + 4D and the angle of esotropia is between 20$^{\Delta}$ and 30$^{\Delta}$. Full amount of hypermetropia should be corrected with early amblyopia therapy.

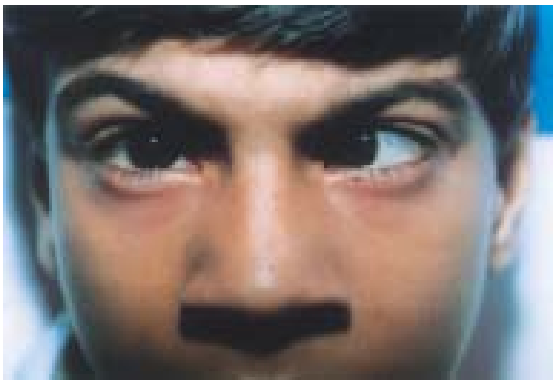


Fig. 23.14A: Esodeviation

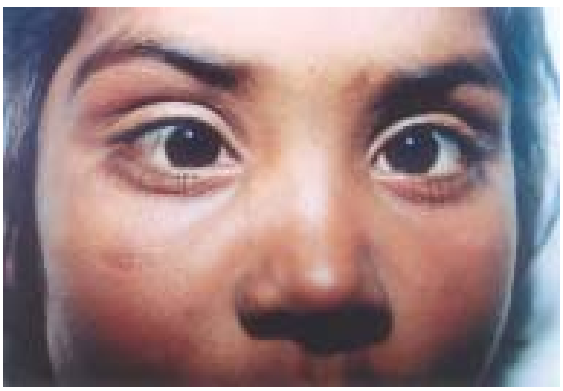


Fig. 23.14B: Accommodative esotropia

b. *Nonrefractive accommodative esotropia* results from an abnormal relationship between accommodative convergence and accommodation (high AC/A ratio). Esotropia develops due to excessive convergence from accommodation. The angle of esotropia is greater for near than for distance. Bifocal correction and surgery are the available management options.
c. *Partially accommodative esotropia* shows a reduction in the angle of esotropia with glasses but residual esotropia persists despite full hypermetropic correction and amblyopia therapy. Surgery may be needed for the correction of residual esotropia.

3. ***Non-accommodative esotropia:*** It develops after 6 months of age and is not associated with accommodative component. It is also known as *basic* or *acquired esotropia*. The treatment of basic esotropia includes amblyopia therapy and surgical correction.
4. ***Secondary esotropia***: Monocular organic lesions like corneal opacity and cataract may cause sensory deprivation esotropia. Overcorrection of exotropia also results in esodeviation.

Exodeviation

The exodeviation (Fig. 23.15) usually manifests at a later age than the esodeviation and is less common than esodeviation. When the sound eye fixates, the deviating eye (which usually has a poor vision) takes the position of rest, which is usually that of divergence.

Exodeviation can be divided into following four types:

1. ***Intermittent exotropia***: It is most common and occurs before the age of 5 years. The deviation is usually larger for distance than for near. The exotropia may be associated with small degree of hypermetropia but amblyopia is uncommon. It has been subdivided into following four groups:
   a. *Basic* shows same deviation for distance and near fixation.
   b. *Divergence excess* shows greater deviation for distance fixation than at near.
   c. *Simulated divergence excess* shows initial greater deviation at distance fixation than at near but becomes same after occlusion of one eye for 1 hour.
   d. *Convergence insufficiency* shows greater deviation at near fixation than at distance. The treatment of intermittent exodeviation includes corrective lenses, unilateral part-time patching, orthoptic exercises and surgery.
2. ***Constant exotropia***: It is a large angle exotropia occurring in older patients as a result of decompensated intermittent exotropia or due to overaction of all the four oblique muscles.
3. ***Congenital exotropia***: It is a large angle constant exotropia occurring before the age of 6 months. It may be associated with neurological and craniofacial anomalies. Early surgery may be of some help.
4. ***Consecutive exotropia***: It occurs following surgical overcorrection of esotropia.

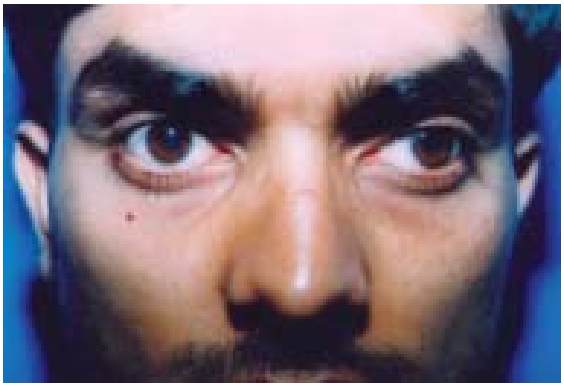


Fig. 23.15: Exodeviation

Vertical Deviations

Vertical deviations of the eye may occur alone or in combination with a horizontal deviation. The vertical deviation is described according to the direction of the vertically deviating nonfixing eye. For example, when the left eye is fixing and the right eye is higher than the left, it is called *right hypertropia*. Vertical deviations are caused by dysfunctional overaction or underaction of the superior and inferior oblique muscles.

A and V-Patterns

Varying degrees of horizontal deviation may occur with upward and downward gaze giving characteristic A and V-patterns.

A-pattern occurs when the horizontal deviation shows a more convergent alignment in upward gaze compared to the downward gaze.

V-pattern occurs when the horizontal deviation shows a more convergent alignment in downward gaze compared to the upward gaze.

Besides the oblique muscle dysfunction, abnormal functioning of horizontal and vertical rectus muscles may contribute to A and V-pattern deviation.

The A and V-patterns are treated surgically. The surgery consists of correction of horizontal deviation, weakening of the inferior oblique muscle for V-pattern correction, and bilateral superior oblique tenotomies for correction of A-pattern.

Clinical Features

Most of the patients of comitant strabismus are symptom-free and brought by their parents for cosmetic purpose. There is no history of diplopia owing to suppression of the image in the deviating eye (development of amblyopia in the squinting eye).

The angle of strabismus remains constant in all directions of gaze. The secondary deviation is always equal to the primary. Ocular movements are full and not restricted in any direction of gaze. False projection is absent and there is no abnormal head posture. These features differentiate the comitant strabismus from the incomitant strabismus (Table 23.4).

Table 23.4: Distinguishing features between comitant and incomitant strabismus

Features	*Incomitant*	*Comitant*
1. History	Non-familial	Familial
2. Onset	Sudden	Insidious
3. Symptoms		
Diplopia	Present	Absent
Nausea and vertigo	Present	Absent
4. Angle of deviation	Changes in certain directions of gaze	Remains constant in all directions of gaze
5. Head and chin position	Abnormal head and chin position to avoid diplopia	Normal position
6. Orientation or projection	Objects are projected too far in the direction of paralyzed muscle	Objects are located at their usual distance
7. Ocular movements	Restricted in the direction of action of paralyzed muscle	Normal in all directions of gaze
8. Secondary changes in extraocular muscles	Contracture or inhibitional palsies are found	No secondary changes are found

Diagnosis

Each case of comitant strabismus should be thoroughly investigated.

Histroy: A careful history must be recorded about the occurrence of strabismus in the family, age of onset of deviation, type and nature of deviation and whether the same eye deviates or it alternates. The history of any preceding illness should be elicited and the progress of the deviation should be noted. On enquiry, it is occasionally revealed that one of the patient's friends, classmates or relations may be suffering from some deviation which the child tends to mimic.

Visual acuity: The visual acuity must be recorded in each case although evaluation of vision in infants is difficult. The *preferential looking test* can be used in infants between the age of 4 months and 6 months. *Tellar acuity card* permits the examiner to observe upon which half of the card the infant fixates. Infants who preferred the striped side have good fixation.

Refractive error: Refractive error, if any, be assessed under full cycloplegia.

Cover/Uncover test: The patient has to fixate on a distant target and one eye is then occluded. At the time of occlusion the fellow eye is observed for any movement. If it moves outwards esotropia is present and if it moves inward it means an exotropia. In bilateral alternate strabismus the cover/uncover test will demonstrate that both eyes fixate and deviate alternately.

Measurement of Angle of Deviation

The angle of deviation must be determined in all cases of comitant strabismus as it is a guide to the management. Several methods are available to assess it.

1. *Corneal reflex test (Hirschberg test):* The test gives a rough idea of the angle of deviation as it estimates the deviation of corneal light reflex from the center of the pupil. A light from a torch is thrown from a distance of 33 cm on the patient's eye, reflexes are normally located at the center of the cornea. When the reflex is situated at the margin of the pupil the deviation is nearly 15°, at the limbus about 45° and midway between the pupillary margin and the limbus about 30° (Fig. 23.16).

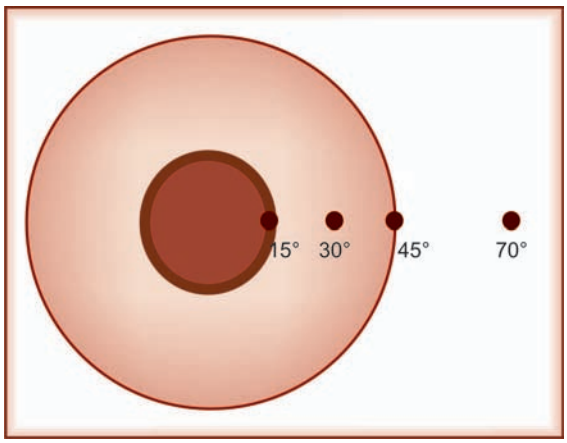


Fig. 23.16: Hirschberg's corneal reflex test

2. *Prism reflex test of Krimsky*: The degree of deviation can also be measured by placing the prisms of increasing powers before the fixing eye until the light reflex is centered in the squinting eye. Alternatively, the prism can be placed in front of the squinting eye until the light reflex is centered on the cornea. In the latter procedure some difficulty is experienced to observe the light reflex under the prism bar (Fig. 23.17).
3. *Measurement on synoptophore*: The angle of deviation is accurately measured on synoptophore. The patient is asked to look through two adjustable tubes and fix at small objects given on appropriate slides. The angle between the tubes is altered until each eye separately attains fixation. This gives the position of the visual axes. Now the corneal reflexes are centred corresponding with the optical axes; the difference between the two represents the angle of deviation.

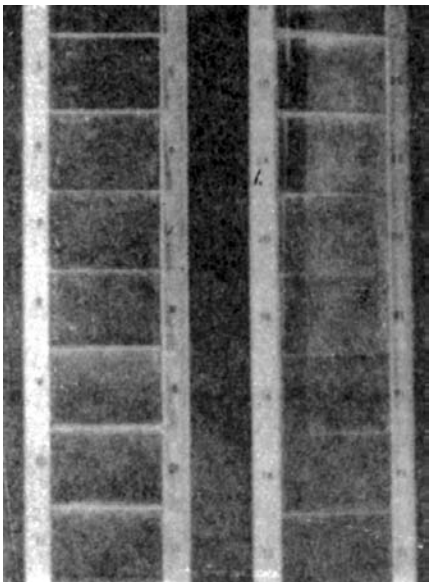


Fig. 23.17: Prism bars

4. *Test for binocular sensory function*: Sensory binocularity can be tested by Worth four dots test and Bagolini test.

Worth four dots test detects the suppression.

The patient wears red (right eye) and green (left eye) glasses and sees a box containing four lights, one white, one red and two green. There can be four possibilities:

1. When 4 lights are seen, it indicates normal fusion and orthophoria. When right eye is dominant, 2 red and 2 green lights are seen. When left eye is dominant, 1 red and 3 green lights are seen.
2. When right eye is suppressed, the patient sees 3 green lights because white light appears green.
3. When left eye is suppressed, the patient sees 2 red lights because white light appears red.
4. In incomitant strabismus with diplopia or rapidly alternating suppression, the patient will see 5 lights simultaneously.

Bagolini after image test is employed for testing retinal correspondence and suppression.

Bagolini striated glasses, when placed in front of the eyes, change the fixation light to an elongated streak. When asked to draw, the patient may draw the position of the perceived images in one of the following patterns.

1. The patients with orthophoria and normal retinal correspondence will draw a diagonal cross.
2. The patient with esotropia will draw 2 lines, vertical line to the left of the horizontal line and in exotropia to the right.
3. The patient with abnormal retinal correspondence with suppression will see only 1 streak.

Determining the Type of Fixation

The fixation can be tested either by a visuoscope or fixation star of an ophthalmoscope. Patient is asked to fixate on the star after closing the fellow eye. In a *central fixation* the image of the star falls on the fovea centralis. In *eccentric fixation* the image may fall on any of the following areas: parafoveal, juxtafoveal, extrafoveal or temporal area, or may remain erratic.

Synoptophore Examination

In addition to the measurement of the angle of strabismus, synoptophore is used to estimate the grades of binocular vision and detection of the normal and the abnormal retinal correspondence.

Treatment

The main principles of strabismus threrapy are:

1. To give a good cosmetic appearance
2. To improve the visual acuity in each eye, and
3. To maintain the binocular vision.

The result of therapy largely depends on the age at which the child is seen. Good cosmetic correction with normal visual acuity is obtained when the therapy is started at six months of age. The ideal objective of treatment is to attain the

binocular vision which is usually difficult to obtain unless the patient had already developed binocularity before the onset of strabismus. The following treatment modalities are adopted:

1. *Correction of Refractive Error*

In all squinting children, a preliminary refraction under full cycloplegia should always be carried out to estimate the error of refraction. A correction of refractive error in the squinting eye is prescribed.

2. *Occlusion*

When the vision is poor in the squinting eye due to disuse, occlusion of the sound eye is advised for 6 weeks. Occlusion is effective upto the age of seven years. Occlusion (patching) should be absolute otherwise the desired improvement is seldom achieved. Sometimes, the deviation is transferred to the occluded eye. It carries a good prognosis and indicates that the vision in the squinting eye was not that poor initially.

In cases of eccentric fixation *inverse occlusion*, i.e., the occlusion of the eccentrically fixing eye, is preferred to prevent stimulation of the abnormal fixing point of the affected eye. Inverse occlusion may be combined with pleoptic treatment. Later when central fixation is restored, conventional occlusion of the sound eye may be done to manage the amblyopia.

Orthoptic Exercises

The cases having a smaller angle of deviation (10°) with good visual acuity but poor binocular vision need orthoptic training for the development of binocular vision of all the three grades. Specially devised exercises on synoptophore are helpful in the development of binocular vision. Orthoptic treatment is a valuable adjunct to the operative management especially in cases with large angle of deviation.

Surgery

Surgical correction of strabismus should be undertaken after obtaining the maximal improvement in visual acuity following patching or penalization. In penalization atropine 1% is used in the normal eye to stimulate the fellow amblyopic eye. Surgical intervention merely provides a cosmetic correction, it does not correct the underlying cause of ocular deviation. The ideal age at which surgery should be undertaken is debatable. However, good results are achieved in those operated between 4 and 6 years of age. There are essentially two surgical approaches for the correction of strabismus, strengthening of a weak muscle (resection) or weakening of an overacting muscle (recession). Free tenotomy or guarded tenotomy is recommended in certain conditions as weakening procedures. Two or more operations may sometimes be required in large angle deviations to straighten the eyes. The type of surgery chosen depends, to a large extent, upon the type of strabismus. Most surgeons prefer to operate upon the deviating eye only in cases of monocular squint except when very large deviation is present. In alternating convergent strabismus, recession of both medial rectus muscles is generally preferred.

NYSTAGMUS

Rapid, regular and rhythmic involuntary movements of the eyes are called *nystagmus*. The movements are usually horizontal, but vertical or rotatory may also occur. The condition is almost always bilateral.

Etiology

Nystagmus has varied etiology. It may be physiological, congenital, infantile or acquired. Physiological nystagmus can be optokinetic, end-point or vestibular.

Optokinetic nystagmus occurs while seeing objects from a moving train or on seeing the alternate black and white strips of a spinning drum (catford drum).

End-point nystagmus can be elicited by asking a person to look in extreme dextroversion or levoversion.

Vestibular nystagmus is seen on stimulation of the tympanic membrane by water (caloric test). A rapid left nystagmus occurs when cold water is poured in the right ear. When warm water is poured in the right ear, rapid right nystagmus occurs.

Nystagmoid jerks may occur in normal persons during fatigue. The movements appear like jerk-type nystagmus and are pronounced on abduction of the eye.

Congenital nystagmus occurs in severely malformed eyes, congenital leukoma and cataract, congenital toxoplasmosis, macular hypoplasia and albinism. It occurs due to nondevelopment of fixation reflex.

Infantile nystagmus occurs in the first year of life. It may be associated with head nodding as seen in spasmus nutans.

Acquired nystagmus occurs in diseases of midbrain, cerebellum and vestibular tract and semicircular canals. It is often seen in multiple sclerosis on extreme lateral gaze. Cerebellar lesions give coarse nystagmus towards the side of the lesion and fine nystagmus on the opposite side.

Miner's nystagmus occurs in coal mine workers. It is characterized by very rapid rotatory ocular movements associated with headache, giddiness and tremors of the head. The incidence of nystagmus is high in workers of poorly illuminated mines suggesting a fixation difficulty as the causative factor.

Labyrinthine nystagmus occurs in diseases of internal ear involving semicircular canals. Destruction of one labyrinth causes nystagmus to the opposite side.

See-saw nystagmus occurs in parachiasmal diseases. It is characterized by elevation and intorsion of one eye while the other eye moves down and extorts.

Convergence-retraction nystagmus is found in Parinaud syndrome (*Sylvian aqueduct syndrome*) which occurs due to the involvement of aqueduct of Sylvius in midbrain glioma or pinealoma. It consists of retraction of eyeball, vertical nystagmus, defective convergence, pupillary abnormality and lid retraction.

Gaze-direction nystagmus occurs in barbiturate intoxication.

Gaze-paretic nystagmus, associated with conjugate gaze weakness, is seen in brainstem lesions at the pontine level.

Up-beat nystagmus occurs in lesions of tegmentum of the brainstem.

Down-beat nystagmus occurs due to lesions of the posterior fossa at the foramen magnum level.

Clinical Features

In congenital and infantile nystagmus the vision is very poor. Besides rhythmic ocular movements, the patient may complain of headache, giddiness, photophobia, moving of stationary objects and occasional diplopia.

Differential Diagnosis

A few ocular motility disorders of childhood like ocular bobbling, flutter-like ocular oscillations, ocular dysmetria and opsoclonus resemble nystagmus. They should be distinguished from nystagmus.

Ocular bobbling is characterized by sudden downward and less commonly upward deviation of eyes; after some time, the eyes slowly come to the primary position. Neoplastic lesions of the pons may cause ocular bobbling.

Flutter-like oscillations of eyes and *ocular dysmetria* signify inability to fixate on an object accurately while shifting the gaze. The eyes overshoot or undershoot the target. They are seen in lesions of the cerebellar connections.

Opsoclonus is marked by wild and chaotic movements of the eyes associated with myoclonic movements of face and extremities. It occurs following encephalitis.

Treatment

Attempts should be made to improve the vision by correction of refractive error of the patient. Prism therapy and large recession of both medial rectus muscles are recommended to transfer the null point (where nystagmus is least) from the eccentric position to straight-ahead position. It also relieves the compensatory head posture.

1. *Correction of refractive error:* Attempts should be made to correct the refractive error by prescribing suitable glasses or fitting of contact lenses. Vision often remains poor in congenital and infantile nystagmus in spite of correction of the error.
2. *Prism therapy:* Prisms are used to correct the head position and shifting the image into the null point area.
3. *Nystagmus surgery*: The surgery improves the visual acuity and broadens the null zone.

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CHAPTER

24 Diseases of the Lids

ANATOMY

Eyelid

The eyeball is protected in the anterior aspect by a set of eyelids, upper and lower. The margins of upper and lower lids meet at the medial and the lateral angle called the *canthi*. They bound an elliptical space, the *palpebral fissure*. There is a semilunar fold of conjunctiva called the *plica semilunaris* at the medial canthus (Fig. 24.1). It represents the nictitating membrane of lower animals. The two eyelids are separated medially by a triangular space, the *lacrimal lake* or *lacus lacrimalis*. A fleshy raised body, the *lacrimal caruncle*, is located in the lacus lacrimalis on the medial side of the plica. Along the plane of the plica lies the *lacrimal papilla* in each lid. On the top of the papilla is the *lacrimal punctum* which leads the tears to the lacrimal canaliculus. The lacrimal punctum divides the lid into ciliary and lacrimal parts. The eyelashes are present only in the ciliary part of the free border of the lid.

Eyelid Margin

The eyelid margin is covered with stratified squamous epithelium which forms the transition between the skin and the conjunctiva. The anterior border of the lid margin is round, while the posterior is sharp and lies closely in contact with the eyeball. This contact facilitates dispersion of tear film and moistening of the cornea and the conjunctiva.

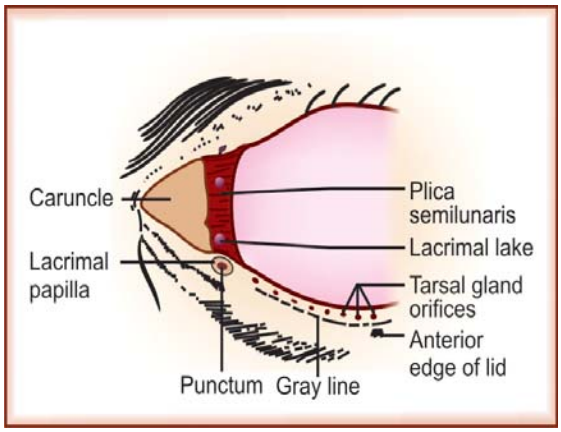

Fig. 24.1: Anatomy of inner canthus

Layers of the Eyelid

Each eyelid from outer to inner aspect has following layers (Fig. 24.2):

1. *Skin* of the lid is delicate and thin and possesses unicellular sebaceous glands. A horizontal fold is present in the skin of the upper lid which becomes prominent on the upward gaze, the superior palpebral sulcus or lid crease. The bulk of the fibers of the levator palpebrae superioris (LPS) muscle pass through the fibers of orbicularis oculi and get inserted into the skin of the upper lid near the superior palpebral

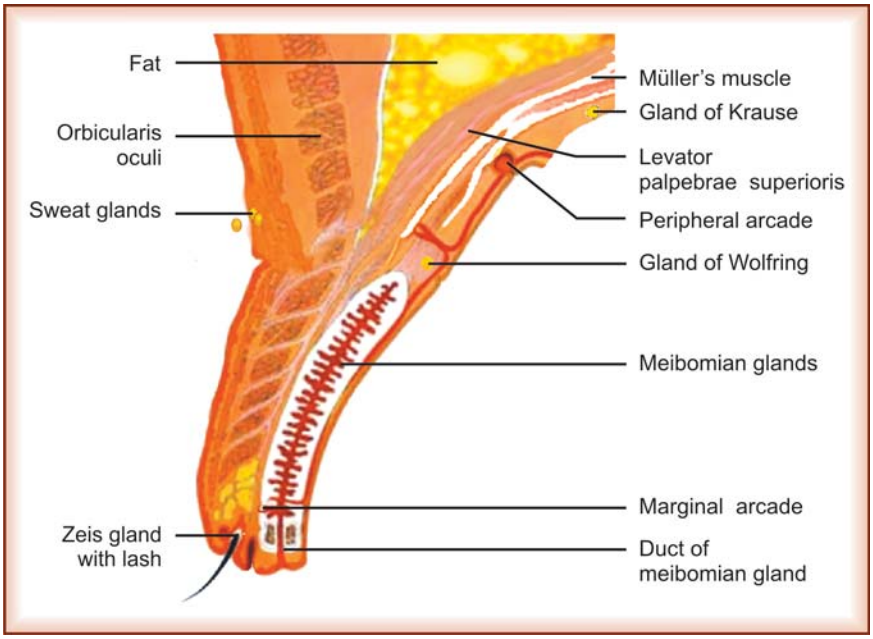

Fig. 24.2: Section through upper lid

sulcus. The skin is loosely attached to the underlying structures; this loose attachment permits easy accumulation of edematous fluid or blood.

2. *Subcutaneous areolar layer* consists of loose connective tissue but no fat.
3. *Layer of striated muscle* is composed of fibers of palpebral part of the orbicularis oculi. It is used for blinking and winking. The orbital part is arranged in a concentric manner around the orbital margin covering the tarsus. It is used for forced closure of the lids. The preseptal portion overlies the orbital septum and the pretarsal portion of orbital part of orbicularis oculi muscle lies anterior to the tarsus. The orbicularis oculi muscle is supplied by the facial nerve.
4. *Submuscular areolar layer* lies deep to the orbicularis oculi. It contains nerves and blood vessels of the eyelid. Anesthetic agents are injected in this layer for lid surgery. An incision at the gray line (a line between the anterior and posterior margins of the lid) splits the lid through the plane into an anterior and a posterior portion.
5. *Fibrous layer* is the main framework of the lid. It has two parts, a thick central part, the tarsal plate and a thin peripheral part, the orbital septum or septum orbitale.

   The tarsal plate maintains the form of the lid and aids to its support. The superior tarsal is larger than the inferior. Some fibers of LPS are attached to the front and lower part of the tarsal plate. They also join the unstriped muscle of Müller at the upper border of the tarsal plate. The meibomian glands (tarsal glands) are embedded in the tarsal plate. The lateral palpebral ligament attaches the lateral ends of the tarsi to the Whitnall tubercle, while the medial ends of the tarsi are attached by the medial palpebral ligament to the lacrimal crest. The septum orbitale is attached to the orbital margin. It is thicker on the lateral side than on the medial and in the upper lid than in the

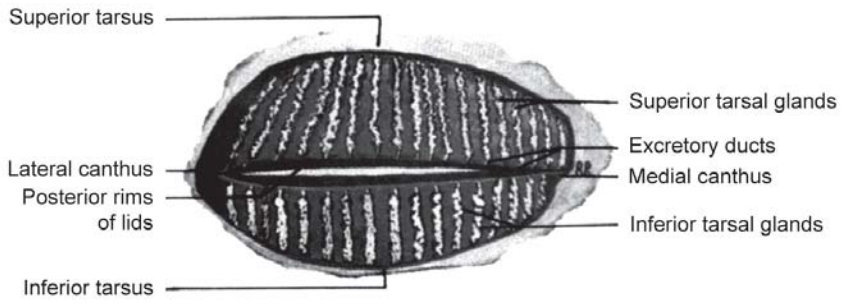

Fig. 24.3: Meibomian glands

lower. In the upper lid the septum is in contact with the orbital fat which separates it from the levator palpebrae superioris.

6. *Smooth muscle fibers of Müller* lie behind the orbital septum. They are believed to help in the retraction of eyeball and elevation of the upper lid. They are supplied by the sympathetic nerves.
7. *Palpebral conjunctiva* covers the posterior part of the lid. It is closely adherent to the tarsus.

Glands of the Eyelid

The glands of Zeis are situated at the lid margin in close association with the cilia. Each gland opens into the follicle of the cilium by a short duct.

The glands of Moll are modified sweat glands and lie between the cilia. The duct of Moll's gland opens either into the duct of Zeis gland or directly into the follicle of cilia.

Meibomian glands (Fig. 24.3) are enormously developed modified sebaceous glands embedded in the tarsi, numbering 30 to 40 in the upper lid and 20 to 30 in the lower. Their ducts open in front of the posterior border of the lid margin. The glands secrete oily secretion which lubricates the eye and prevents evaporation of tears from the cornea.

Blood Supply of the Eyelid

The eyelids are supplied by the medial and lateral palpebral arteries, branches of the ophthalmic and lacrimal arteries respectively. They form two main arcades: the superior peripheral arcade lying between the upper part of the tarsus and the orbicularis, and the inferior peripheral arcade, nearly 3 mm above the lid margin (Fig. 24.4). The superior arcade is reinforced by the branches from the superficial temporal, lacrimal and supraorbital arteries, while the inferior by the transverse facial and facial arteries.

Each lid is drained by the pretarsal and the posttarsal plexus into the subcutaneous and ophthalmic veins, respectively. Lymphatics from most of the upper lid and lateral half of the lower lid drain into the preauricular lymph nodes and the medial portions of both the lids drain into the submandibular lymph nodes.

Nerve Supply of the Eyelid

The orbicularis oculi is supplied by the facial nerve, the LPS by the upper division of the oculomotor nerve and the Müller's muscle by the sympathetic nerves. The sensory supply to the upper lid comes from the ophthalmic division of the trigeminal nerve. The lower lid is supplied through the infraorbital nerve, a branch of the maxillary division of the trigeminal nerve.

CONGENITAL ANOMALIES OF THE LIDS

Ablepharon is an extremely rare congenital defect wherein the lid is not developed.

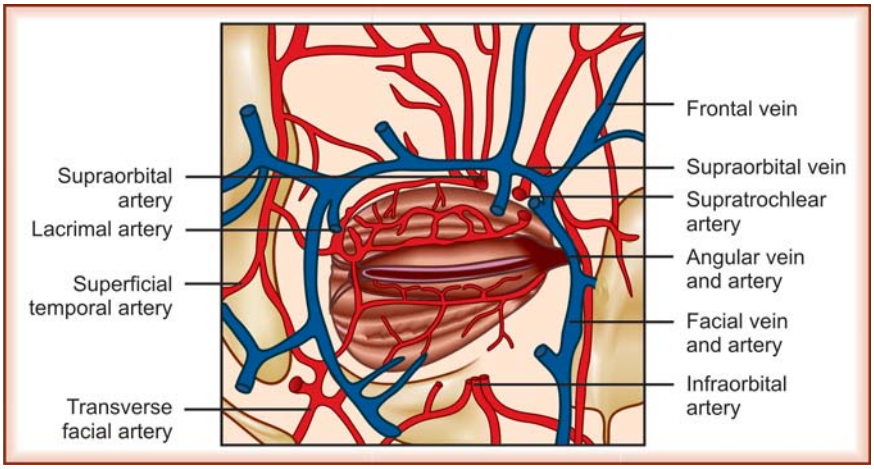

Fig. 24.4: Blood supply of the lid

Microblepharon is a rare anomaly in which the lids are abnormally small.

Cryptophthalmos is a rare anomaly in which a fold of skin passes from the eyebrow over the malformedeye to the cheek.

Ptosis is drooping of the lid and a fairly common congenital defect.

Epicanthus is frequently bilateral and often associated with ptosis (Fig. 24.5). A semilunar fold of skin is present above and sometimes covers the inner canthus. It gives an appearance of pseudoconvergent strabismus and disappears as the nose develops. It is usually found in Mongolian races. The anomaly can be corrected by plastic surgery.

Distichiasis is a congenital anomaly of eyelashes wherein an extra posterior row of cilia is present. This additional row of the lashes occupies the position of meibomian glands which are reduced to ordinary sebaceous glands. Distichiasis causes irritation of the eye. It can be managed by cryo application to the posterior lid margin after splitting the eyelid along the gray line.

Coloboma of the lid (Fig. 24.6) is characterized by a notch that is usually situated at the junction of the middle-third and the medial-third of the upper lid. It may be associated with the presence of dermoid. Incomplete closure of the embryonic facial cleft and pressure of the amniotic bands are implicated in the etiology of the coloboma. The coloboma requires a plastic repair. It can be reconstructed by a graft from the lower lid.

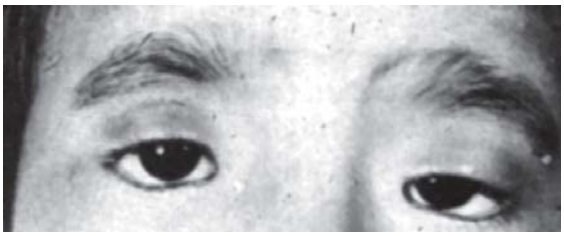

Fig. 24.5: Ptosis of left eye with epicanthus

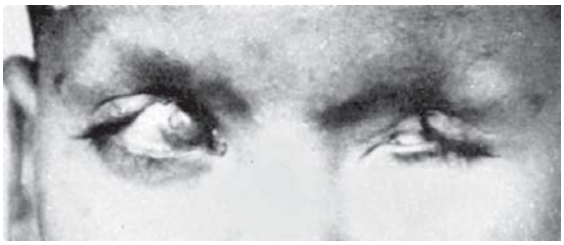

Fig. 24.6: Colobomas of both upper lids

EDEMA OF THE LIDS

Edema of the lids is common due to looseness of the tissues. It occurs either as an inflammatory edema or a passive edema.

Inflammatory edema is often found in acute conjunctivitis, tarsitis, dacryocystitis and orbital cellulitis. Acute painless edema of the lid has an allergic basis. It may be found in drug (atropine) allergy or angioneurotic edema (Fig. 24.7).

Passive edema of the eyelid is a common feature of cavernous sinus thrombosis, congestive heart failure, nephrotic syndrome, hypoproteinemia and anemia.

INFLAMMATION OF THE LIDS

The lids are often involved in sensitization reaction to cosmetic dyes and drugs. Atropine reaction is a typical example. Dermatitis or eczema may spread to the lids. Herpes zoster and anthrax vesicles may erupt on the lids. Syphilitic affections of the lids are uncommon. A primary chancre may develop following a kiss. Gummata or syphilitic tarsitis may cause thickening of the tarsus. A pustule on the lid may develop as a result of inoculation from the recently vaccinated arm of a baby.

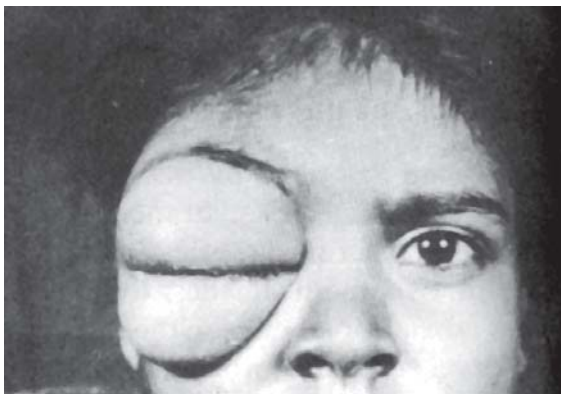

Fig. 24.7: Angioneurotic edema of right upper and lower lids

Blepharitis

Blepharitis is a chronic inflammation of the lid margin. Clinically, it occurs in two forms, squamous blepharitis and ulcerative blepharitis.

Squamous Blepharitis

Squamous blepharitis is characterized by the presence of small white scales at the root of the lashes (Fig. 24.8) which may fall out but are replaced without distortion.

Etiology

The blepharitis is common in children who suffer from dandruff of the scalp. The squamous blepharitis is essentially a metabolic disorder and often associated with seborrhea. Perhaps, certain organisms produce lipolytic co-enzymes that split the neutral lipids into free fatty acids which irritate the lid margins and the conjunctiva.

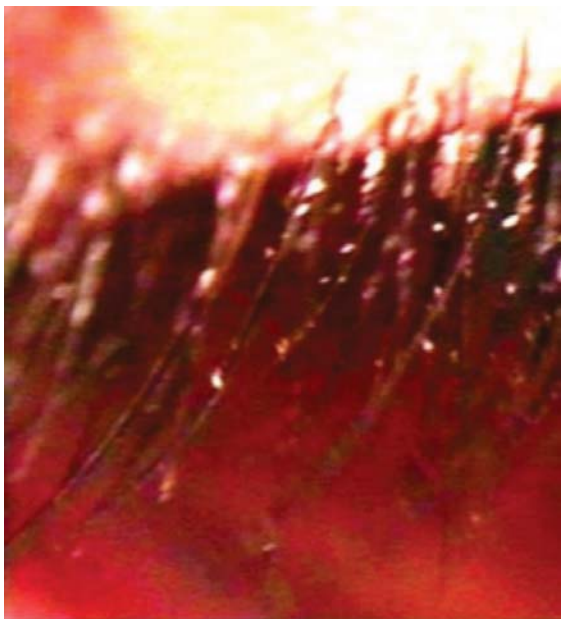

Fig. 24.8: Squamous blepharitis (*Courtesy*: Dr Vinay Agrawal, Mumbai)

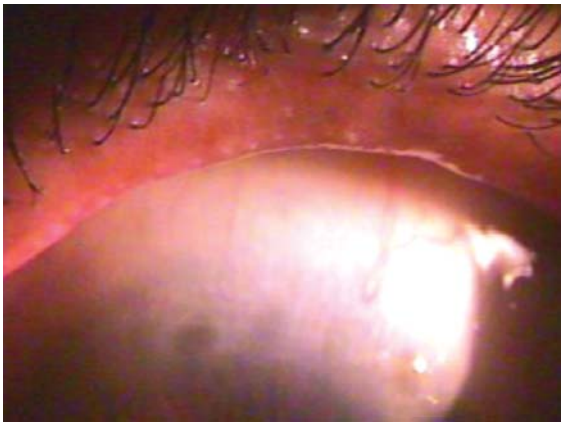

Fig. 24.9: Squamous blepharitis associated wih meibomianitis

Clinical Features

Irritation, itching and watering are common symptoms. White dandruff-like scales on the eyelid margins are often found. If the scales are removed the underlying area is found to be hyperemic but not ulcerated. Squamous blepharitis is often associated with meibomianitis (Fig. 24.9).

Complications

Squamous blepharitis rarely causes complications, but constant irritation due to free fatty acids may lead to chronic papillary conjunctivitis and punctate epithelial erosions of the cornea.

Ulcerative Blepharitis

Ulcerative blepharitis is an infective condition of the lid margin characterized by the deposition of yellow crusts at the roots of eyelashes, swelling of the lid margins and falling of the eyelashes (Fig. 24.10). The removal of the crust leaves a small round ulcer which bleeds readily.

Etiology

Ulcerative blepharitis is common in debilitated children with poor personal hygiene. *Staphylococcus aureus* (coagulase positive) is the most common organism causing ulcerative blepharitis. The condition may be secondary to chronic conjunctivitis. Eyestrain and refractive errors are known risk factors. Certain parasites such as crab louse and *Demodex folliculorum* can cause the disease.

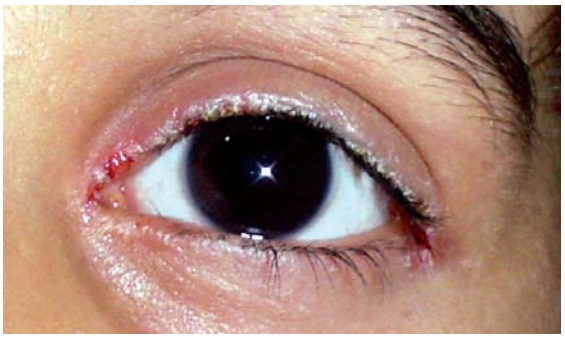

Fig. 24.10: Ulcerative blepharitis (*Courtesy*: Prof. Manoj Shukla and Dr Prashant Shukla AMUIO, Aligarh)

Clinical Features

Intense itching, swelling and redness of lid margins, falling of lashes, watering and photophobia are common symptoms of ulcerative blepharitis. The crusts glue the eyelashes together and cause difficulty in opening the lids. The ulcerative blepharitis has a chronic course and is often accompanied with chronic conjunctivitis, the two together are known as *blepharoconjunctivitis*. The presence of nits at the roots of eyelashes is diagnostic of parasitic blepharitis.

Complications

When blepharitis persists for a long time, the ulcerative process extends deeply and destroys the hair follicles resulting in falling of cilia. The cilia are often not replaced, and if they regrow only a few distorted ones come. The condition is known as *madarosis* (Fig. 24.11). The shallow ulcers of blepharitis heal by fibrosis, the contraction of fibrous tissue may cause misdirection of a few eyelashes which rub against the cornea, the

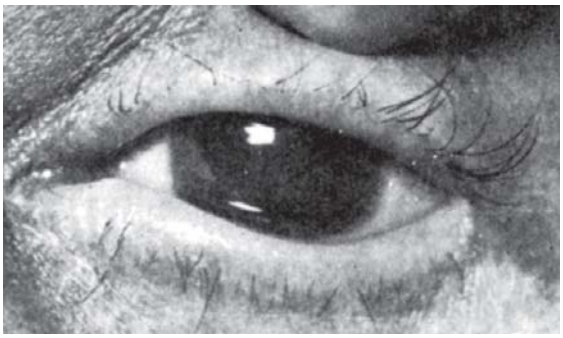

Fig. 24.11: Madarosis

condition is termed as *trichiasis*. The excessive fibrosis of the lid margin leads to its thickening known as *tylosis*.

Occasionally the contraction of the scar tissue pulls the conjunctiva of the lower lid making the acute posterior border more or less rounded so that its capillary action is disturbed resulting in epiphora. The constant moistening of the skin leads to eczema of the lower eyelid which eventually causes *ectropion*. The ulcerative blepharitis may also be associated with recurrent styes and angular conjunctivitis. The staphylococcal exotoxins may induce *punctate epithelial erosions* predominantly in the lower part of the cornea, *catarrhal marginal corneal ulcers* and, rarely, *phlyctenulosis*.

Basic differences in the clinical features of squamous and ulcerative blepharitis are listed in Table 24.1.

Treatment

Treatment of blepharitis consists of proper removal of the crust, application of specific antibiotic ointment depending on the sensitivity of the organism and massage of the lid margin so that the drug may reach the follicles of eyelashes where organisms lie hidden. The crust can be softened and removed through bathing with warm 3% sodium bicarbonate lotion or hydrogen peroxide.

Mechanical expression of the meibomian glands is particularly helpful in controlling squamous blepharitis because the procedure reduces the vicarious secretion from the glands. Associated seborrheic dermatitis and dandruff need special care and should be managed by medicated shampoo. Many eyes with blepharitis show a rapid tear film break-up time (BUT) suggesting dryness (sicca). They show symptomatic relief if treated with tear substitutes.

Table 24.1: Differentiating features of squamous and ulcerative blepharitis

Features	*Squamous*	*Ulcerative*
1. Etiology	Seborrhea	Staphylococcus
2. Deposits on eyelashes	Shiny waxy	Brittle scales/ulcers
3. Removal of deposits leaves	Hyperemic zone	Bleeding ulcers
4. Hair follicles	Not destroyed	Destroyed
5. Meibomian secretion	Vicarious	Normal
6. Eyelashes	Normal	Trichiasis, madarosis and poliosis
7. Lid margin may present	Chalazion	Stye
8. Conjunctiva	Mild conjunctivitis	Severe conjunctivitis
9. Dryness	Absent	Present
10. Corneal complications		
a. Punctate epithelial erosions	May be seen	Commonly seen
b. Marginal ulcers	Absent	Present

The application of an antibiotic ointment and massage should be done 2 to 3 times a day and continued for more than 2 to 3 weeks after obtaining the relief.

Systemic tetracycline administration is found to be useful in patients with squamous blepharitis. Systemic corticosteroids should be avoided in ulcerative blepharitis. However, topical corticosteroids are very effective in the management of papillary conjunctivitis and marginal keratitis as they control the hypersensitivity reaction.

Parasitic blepharitis is treated by removal of nits by forceps and delousing of the patient.

INFLAMMATION OF THE GLANDS OF THE LIDS

Stye (Hordeolum Externum)

Hordeolum externum is a suppurative inflammation of one of the Zeis glands.

Stye is common in young adults with refractive errors and muscular imbalance and may occur in crops. It is often caused by *Staphylococcus aureus*.

Clinical Features

A painful swelling appears on the lid margin. The inflamed gland becomes hard, swollen and tender, and soon an abscess forms.

Treatment

Hot compresses are beneficial in the initial stages of the stye, but when the abscess is formed it should be evacuated by a small incision. Application of antibiotic ointment prevents recurrences. If crops of styes occur, diabetes mellitus must be excluded and a course of systemic antibiotics may be prescribed. Staphylococcal vaccine is recommended in recurrent styes.

Hordeolum Internum

Hordeolum internum is a suppurative inflammation of the meibomian gland which can occur due to secondary infection of a chalazion.

It is less frequent but a more violent inflammation than stye. The gland is enlarged and causes enormous swelling of the lid (Fig. 24.12). Occasionally, it may burst through the duct or the conjunctiva. An incision may be required to evacuate the pus.

Chalazion

Chalazion (Fig. 24.13) is a chronic granulomatous inflammation of the meibomian gland.

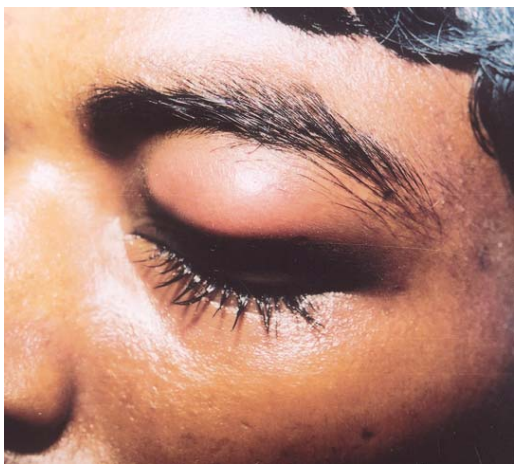

Fig. 24.12: Hordeolum internum

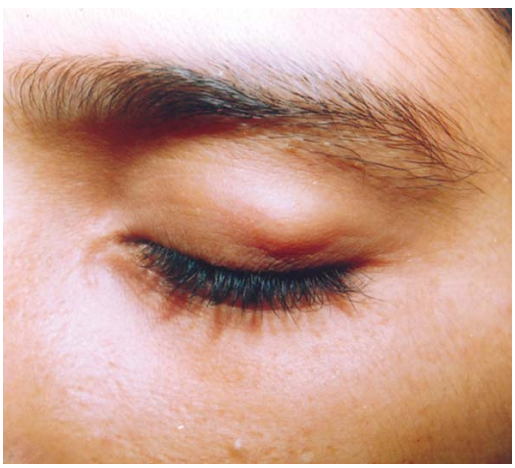

Fig. 24.13: Chalazion of left upper lid

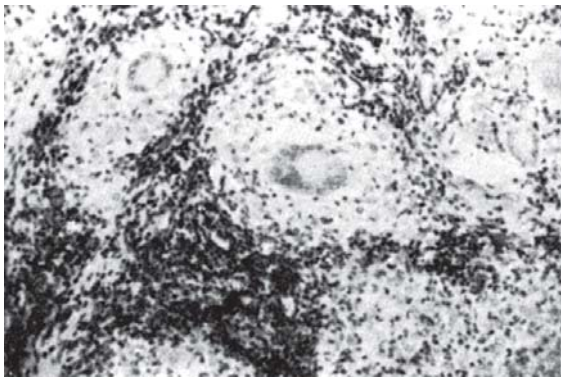

Fig. 24.14: Histopathology of chalazion

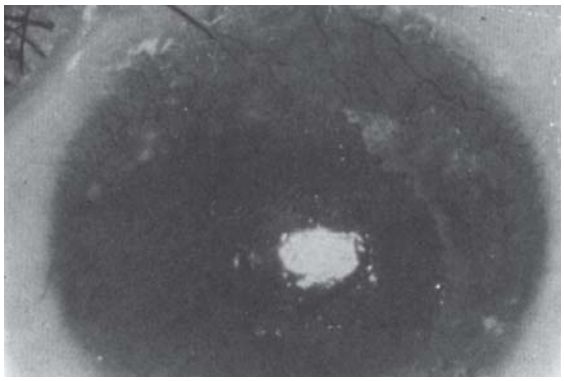

Fig. 24.15: Trichiasis with corneal vascularization

Etiology

Chalazion is commonly seen in adults and often occurs in crops. It perhaps develops as a result of infection by an organism of low virulence or by chronic irritation. The entire gland is replaced by a granulation tissue containing giant cells (Fig. 24.14).

Clinical Features

Chalazion is a painless, round, smooth swelling which can be palpated by passing a finger over the lid. There may be more than one chalazion. On eversion of the lid, the palpebral conjunctiva is red or gray over the chalazion.

Spontaneous resolution of a chalazion seldom occurs. It may form a granuloma following its extrusion through the conjunctiva or rarely through the duct of the gland (*marginal chalazion*).

Treatment

The chalazion requires incision and curettage. Conjunctival approach is often preferred. After a proper local anesthesia a chalazion clamp is applied and the lid is everted. A vertical incision is made at the site of discoloration in the conjunctiva. The contents are thoroughly scraped with a chalazion scoop. Intralesional injection of triamcinolone acetonide may resolve a small chalazion.

A recurring chalazion in old age should arouse the suspicion of a malignancy. Such a case needs meticulous dissection of chalazion and histopathological examination to rule out adenocarcinoma of the meibomian gland.

DEFORMITIES OF THE LID MARGIN AND PALPEBRAL APERTURE

Trichiasis

Trichiasis is a condition where a few cilia are misdirected backwards and they rub against the cornea.

Etiology

Trichiasis is caused by trachoma, ulcerative blepharitis, ocular burns, membranous conjunctivitis, injury or an operation on the lid margin. It is also seen in congenital distichiasis.

Clinical Features

Foreign body sensation or irritation, lacrimation, photophobia and pain are common symptoms of trichiasis. The misdirected lashes may rub against the cornea or cause corneal erosions and vascularization (Fig. 24.15).

Treatment

The condition may be dealt with removal of misdirected eyelashes by epilation or the hair follicle can be destroyed by electrolysis or diathermy.

Electrolysis is a procedure in which the follicle of eyelash is destroyed by passage of an electric current (3 to 5 milliamperes) through a fine needle inserted into the root of the eyelash. Then the eyelash is removed with the help of a forceps. It is a painful procedure and needs multiple sittings.

The hair follicles can also be destroyed by passage of a current of 30 milliampere for 10 seconds through a diathermy needle.

Cryotherapy is an effective method for destroying the misdirected lashes. The cryo probe with –20° C temperature is applied onto the root of the eyelash, a freeze-thaw-freeze technique is used. It destroys the hair follicle, but leaves a depigmented area after healing. Trichiasis can also be corrected with argon laser but it is less effective than cyotherapy. Surgery may be needed in some cases.

Entropion

Entropion (Fig. 24.16) is a condition wherein the lid margin is rolled inwards. The inturned eyelashes rub against the cornea and the conjunctiva and cause irritation, watering and photophobia.

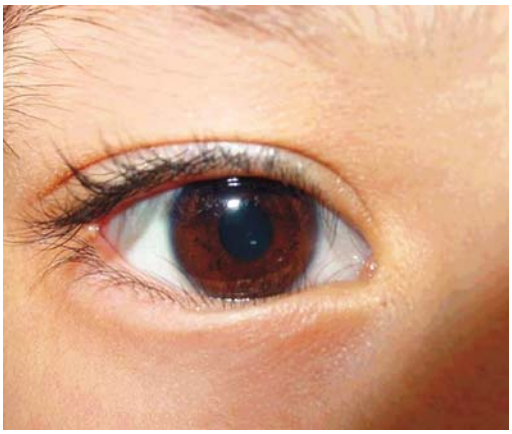

Fig. 24.16: Entropion of lower lid (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

Types of Entropion

Entopion may occur in 4 forms:

1. Spastic
2. Cicatricial
3. Involutional, and
4. Congenital.

1. *Spastic entropion*: Spastic entropion occurs due to the spasm of orbicularis oculi, particularly when the eyeball is deeply set, small (microphthalmos) or absent. The condition may occur in old age owing to the atrophy of orbital fat or after prolonged tight bandaging. Spastic entropion is almost always seen in the lower lid.
2. *Cicatricial entropion*: Cicatricial entropion frequently occurs in the upper lid and is caused by the contraction of the conjunctival scar associated with distortion of the tarsal plate as found in trachoma, membranous conjunctivitis, chemical burns, trauma and Stevens-Johnson syndrome.
3. *Involutional entropion* (senile): The senile entropion usually occurs in the lower eyelid. It is caused by a number of factors such as horizontal laxity of the eyelid, disinsertion of eyelid retractors and overriding of the preseptal orbicularis oculi muscle.
4. *Congenital entropion*: It is rare and often associated with microphthalmos and needs repair.

Clinical Features

Entropion gives almost all the symptoms of trichiasis. The entropion can be of 3 grades. It may be *mild*, when only posterior lid border inturns, *moderate*, when the intermarginal strip rotates inwards, and *severe*, when entire lid margin rolls inwards.

Complications of entropion include corneal ulcer and corneal vascularization.

Treatment

Spastic entropion due to bandaging can be relieved by discarding the bandage. Persistent spastic entropion may need an injection of 1 ml of 80% alcohol subcutaneously along the margin of the lid. Sustained relief can be obtained by skin-muscle operation in which a piece of skin along with the underlying strip of orbicularis oculi is removed.

Cicatricial entropion usully needs surgical repair. A number of operations to correct entropion of various grades are devised, the common ones are described in the chapter on *Operations on Eyeball and Adnexa*.

The procedures to repair the involutional entropion address to the cause. These include horizontal eyelid tightening and repair of retractors.

Ectropion

Rolling out of the lid margin is called *ectropion.*

Types

Ectropion occurs in five forms:

1. Spastic
2. Cicatricial
3. Senile
4. Paralytic, and
5. Mechanical.

Spastic ectropion: Spastic ectropion occurs from a powerful contraction of orbicularis oculi (blepharospasm) when the lids are well supported by a prominent globe or when they are relatively short. It usually occurs in children, particularly during an attempt to examine a sore eye.

Cicatricial ectropion: It results from the destruction of the skin of the lid by burn (Fig. 24.17), ulcers, trauma, gangrene, chronic conjunctivitis or blepharitis. Occasionally, oesteomyelitis of the orbital bones may produce a cicatricial ectropion.

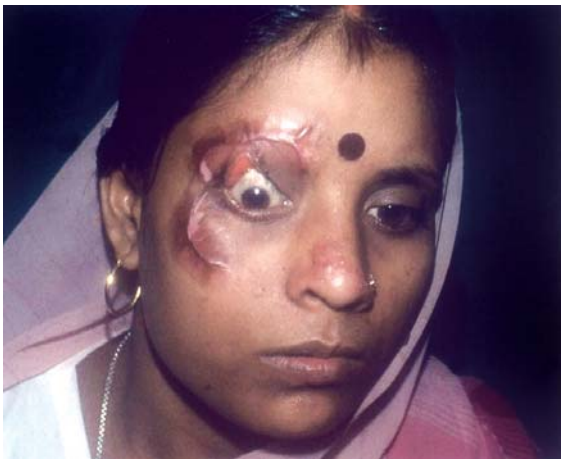

Fig. 24.17: Ectropion of upper lid following burn of face and neck (*Courtesy:* Prof. V Bhattacharya, IMS, BHU, Varanasi)

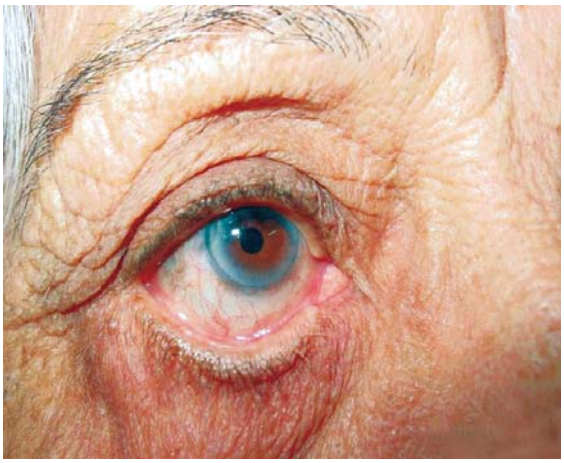

Fig. 24.18: Senile ectropion of lower lid (*Courtesy:* Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

Senile (involutional) ectropion: A laxity of the tissue of the lower lid or loss of tone of orbicularis oculi may develop in old age resulting in senile ectropion (Fig. 24.18).

Paralytic ectropion: Paralytic ectropion follows the seventh cranial nerve paralysis/palsy (Bell's palsy). Road traffic accident, intracranial surgery and middle ear disease may implicate the facial nerve.

Mechanical ectropion: It is caused by the weight of a tumor or granuloma of the lower eyelid.

Clinical Features

Like entropion, ectropion can also be divided in 3 grades: *mild*, when only the punctum is everted, *moderate*, when palpebral conjunctiva is visible due to eversion of the eyelid margin, and *severe*, when the lower fornix is exposed.

Annoying epiphora due to eversion of the lower punctum and exposure conjunctivitis due to eversion of the lower lid margin are common. The exposed conjunctiva becomes dry, thickened and discolored. Exposure keratitis may also occur.

Treatment

The spastic ectropion can be managed by treating the cause of blepharospasm or by applying a well fitting bandage. Epiphora can be corrected by cauterization just posterior to the punctum or slitting the canaliculus. However, most of the other types of ectropion need plastic operations.

Senile ectropion can be corrected by following surgical procedures:

1. *Medial conjunctivoplasty:* It is an effective procedure to manage epiphora. A horizontal fusiform piece of conjunctiva (7 mm long and 4 mm high) nearly 4 mm interior to the punctum is excised and sutured to its margins.
2. *Bick's procedure:* A full-thickness shortening of the lower eyelid just medial to the lateral canthus is recommended in moderate cases of ectropion.

Cicatricial ectropion often needs plastic repair. The pull caused by the scar on the lower eyelid can be corrected by V-Y plasty or Z-plasty (*Elschnig's operation*).

Byron Smith modification of Kuhnt-Szymanowski procedure is recommended in severe and marked cases of ectropion especially of the lateral half of the lower eyelid. The procedure includes shortening of the eyelid with its temporal margin elevation.

Paralytic ectropion needs lateral tarsorrhaphy. The palpebral aperture is shortened by a temporary or a permanent lateral tarsorrhaphy.

Symblepharon

Symblepharon is a condition where adhesion develops between the eyelid and the eyeball.

Etiology

Symblepharon occurs due to membranous conjunctivitis, chemical burns, ulcers, trauma, ocular pemphigus and ocular surgeries. Bands of fibrous tissue are formed between the raw areas in the palpebral conjunctiva and the globe.

Types

Depending on the site of development of adhesion, the symblepharon can be of 3 types: anterior, posterior and total.

In *anterior symblepharon* adhesion occurs between the lid and the bulbar conjunctiva or the cornea (Fig. 24.19).

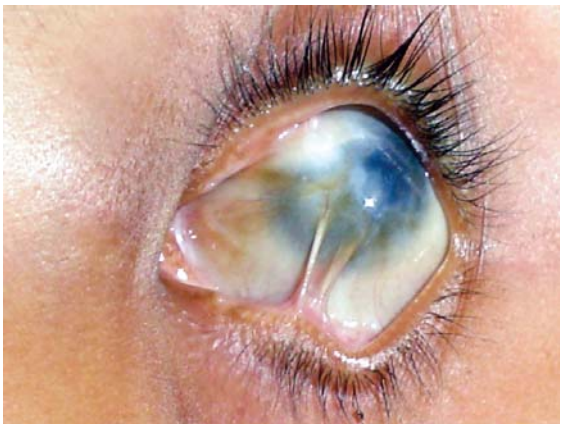

Fig. 24.19: Anterior symblepharon (*Courtesy:* Prof. Manoj Shukla and Dr Prashant Shukla, AMUIO, Aligarh)

In *posterior symblepharon* the upper or the lower or both fornices are obliterated.

In *total symblepharon* the lids get completely adherent to the globe.

Clinical Features

Limitation of ocular movements, diplopia, improper closure of the lids (lagophthalmos) and disfigurement are common features of symblepharon.

Treatment

The symblepharon can be prevented by efficient and adequate treatment of membranous conjunctivitis, wearing of contact shell following burn and meticulous suturing of the wound of the conjunctiva. Bands once formed need excision and the raw surfaces should be covered with conjunctival or buccal mucosal grafts or amniotic membrane.

Ankyloblepharon

Ankyloblepharon is the adhesion of the margins of the upper and the lower eyelid. It may be either a congenital or an acquired condition (following burns). The adhesions can be separated by excision.

Blepharophimosis

Blepharophimosis is the narrowing of the palpebral aperture. The condition is often congenital, but sometimes the aperture appears to be small owing to the presence of a vertical fold of skin at the outer canthus**.** Blepharophimosis can be managed by canthoplasty.

Blepharospasm

Blepharospasm is defined as a forceful closure of the eyelids.

Types

Blepharospasm is of two types: (i) essential blepharospasm, and (ii) reflex blepharospasm.

Essential Blepharospasm

Etiology

The essential blepharospasm affects women more frequently than men. The age of onset is between 45 and 65 years. The cause of blepharospasm is not known, it may be of central origin (basal ganglion).

Clinical Features

The condition starts as an increased blinking and mild twitching of the lids and progresses to forced contractions of the eyelids. Inability to open the lids may limit the patient's routine activities. Spasmodic closure of lids may result in spastic ectropion in children and entropion in elderly persons.

Treatment

The treatment of essential blepharospasm includes following procedures:

1. Repeated periodic injections of botulinum toxin A (Botox) are effective in temporary (3-4 months) chemical denervation and muscle paralysis.
2. Unresponsive patients may need surgical myectomy of the orbicularis oculi fibers.
3. Surgical ablation of the facial nerve, although recommended, is not preferred.

Reflex Blepharospasm

Etiology

The reflex blepharospasm is commonly caused by bright light, acute conjunctivitis, corneal ulcer and keratitis. Severe dry eye syndrome can also lead to contraction of the periocular musculature.

Treatment

Reflex blepharospasm can be managed by lubricants, removal of the sensory stimulus and sedatives.

Lagophthalmos

Lagophthalmos (Fig. 24.20) is a condition in which the palpebral aperture cannot be closed voluntarily.

Etiology

Lagophthalmos often occurs due to the paralysis of orbicularis oculi (Bell's palsy), cicatricial ectropion and proptosis (owing to orbital tumor or thyrotoxicosis). The patients who are extremely ill or comatosed fail to close their eyes.

Clinical Features

Non-closure results in the development of exposure keratitis that is usually seen in the lower part of the cornea.

Treatment

Frequent applications of lubricating ointment and instillation of artificial tears keep the cornea moist. Use of soft bandage contact lenses and partial or total tarsorrhaphy may prevent exposure keratitis.

Ptosis (Blepharoptosis)

The term ptosis, or more accurately blepharoptosis, refers to drooping of the upper eyelid.

Classification

Ptosis may be classified according to the time of onset (congenital and acquired) or the underlying etiology. Etiological classification is more useful and includes following types of ptosis:

1. Myogenic
2. Aponeurotic
3. Neurogenic
4. Mechanical, and
5. Traumatic.

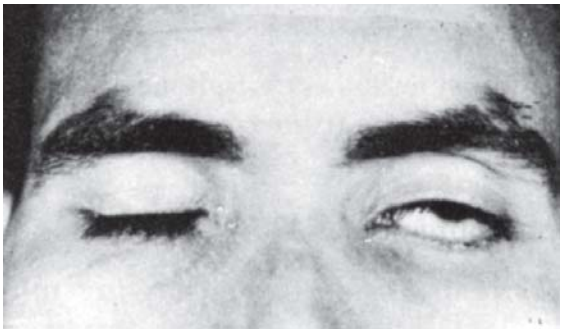

Fig. 24.20: Lagophthalmos of left eye

Myogenic ptosis is caused by the maldevelopment of levator palpebrae superioris. The normal muscle fibers are replaced by fibrous or adipose tissue resulting in diminished contraction and relaxation of the muscle. Myogenic ptosis may be associated with improper development of the superior rectus muscle.

Acquired myogenic ptosis is relatively uncommon and may be seen in myasthenia gravis or muscular dystrophy.

Myasthenia gravis is characterized by a generalized muscular weakness and rapidly developing fatigue of muscles owing to destruction of acetylcholine receptors at the post-synaptic membrane.

Myasthenia gravis causes a bilateral asymmetrical ptosis usually marked towards the end of the day when the patient is tired. Ptosis and convergence deficiency are the early and prominent ocular features. The degree of ptosis increases on prolonged fixation or on elevation. Twitching of the upper lid (*twitch sign of Cogan*) may be found when the patient quickly redirects his gaze from the downward to the primary position. The extraocular muscle weakness causes diplopia in about 50% of cases, but pupillary muscles are seldom affected.

Injection of prostigmine or intravenous tensilon (anticholinesterase) provides a temporary but rapid improvement in muscle action due to the accumulation of acetylcholine at the neuromuscular junction. Thus, it forms a basis for the diagnostic test.

Ptosis associated with the weakness of the facial muscles is found in dystrophia myotonica.

Slowly progressive ptosis is an important feature of ocular myopathy (progressive external ophthalmoplegia).

Aponeurotic ptosis is the most common form of acquired ptosis and caused by stretching and disinsertion of the levator aponeurosis owing to frequent rubbing of the eyes, wearing rigid gas permeable contact lenses, and traction during ocular surgery. In the aponeurotic ptosis, the LPS function is usually normal.

Neurogenic ptosis is caused by innervational defects that occur during embryogenesis. It is frequently associated with congenital oculomotor nerve palsy, Horner's syndrome and Marcus Gunn jaw-winking syndrome.

Mechanical ptosis is caused by excessive weight of the upper lid. It may be due to multiple chalazia, neurofibromatosis, excessive cicatrization of tarsal plate in trachoma, and benign and malignant tumors of the upper lid.

Traumatic ptosis is frequent and is caused by trauma to the LPS muscle or its aponeurosis. Orbital and neurosurgical procedures may lead to ptosis.

Pseudoptosis

Pseudoptosis is an apparent drooping of the upper eyelid often seen in anophthalmos, microphthalmos, enophthalmos, phthisis bulbi and dermatochalasis. Pseudoptosis must be differentiated from the true ptosis.

Work Up

History

The most common form of ptosis is congenital. It may be unilateral or bilateral, partial or complete (Figs 24.21 and 24.22). The condition is usually hereditary. A complete clinical history in all cases of congenital ptosis should be recorded to exclude other causes of ptosis such as trauma or neurological disorders. The presence of congenital ptosis could be confirmed by photographs of the patient taken earlier.

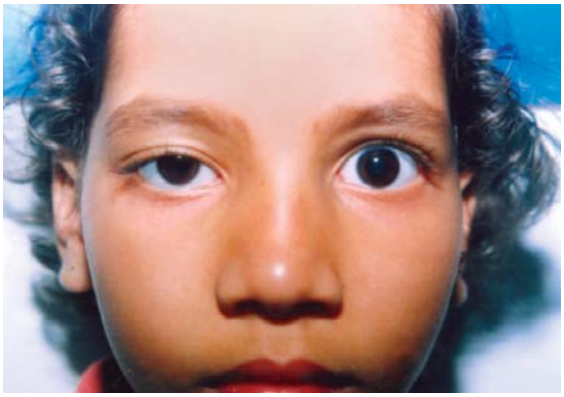

Fig. 24.21: Partial unilateral ptosis of right eye

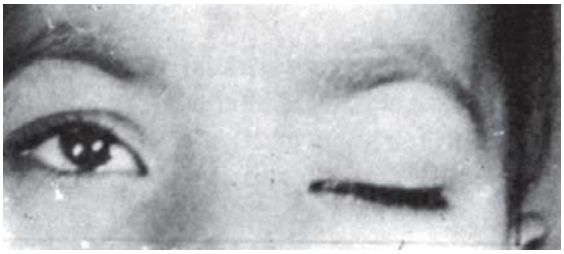

Fig. 24.22: Complete unilateral ptosis of left eye

Assesssment

Proper assessment of ptosis needs following measurements:

1. *Vertical interpalpebral fissure height* is measured between the upper eyelid and the lower eyelid while the patient is fixing at a distant object.
2. *Margin reflex distance (MRD)* is the distance between the upper eyelid margin and the corneal light reflex in the primary position of gaze. MRD is the most important measurement in assessing the degree of ptosis. In the primary gaze, the upper 2 mm of the cornea is usually covered by the upper eyelid.

In unilateral ptosis, the difference between the MRD of two eyes can determine the degree of ptosis. However, in bilateral ptosis, the extent of cornea covered by the ptotic lid is measured by a scale and then 2 mm is subtracted from it.

Ptosis is graded as:

Mild: about 2 mm

Moderate: about 3 mm

Severe: > 3 mm.

3. *Levator function*: The LPS action can be measured by a scale. The examiner places the thumb against the patient's eyebrow. The patient is advised to look down and the reading on the scale opposite the lid margin is noted. Then the thumb is pressed firmly to negate the frontalis action and the patient is asked to look upwards. The position of lid margin against the scale is noted again and the difference between the two readings gives the LPS function. The levator function may be:

   Good: 8 mm or more

   Fair: 5 to 7 mm

   Poor: 4 mm or less.

   In mild ptosis, there is some weakness of LPS, while in severe ptosis there is a near complete inaction of the muscle. In severe degree of ptosis, the lid droops down covering the pupil and thus interferes with the vision.
4. *Upper eyelid crease:* The crease is formed by the insertion of the levator muscle fibers into the skin. A high, duplicated or asymmetric crease suggests abnormal insertion of the levator aponeurosis.

Investigations

Besides the above mentioned measurements, other examinations in a case of ptosis include:

1. Checking the head position and chin elevation, and associated weakness of extraocular muscles especially the superior rectus.
2. Presence of lagophthalmos and the quantity and quality of the tear film.
3. Presence of upward and outward rolling of the eyeball on forced closure of the eyelids (*Bell's phenomenon*).
4. Corneal sensation.
5. Variations in the amount of ptosis on jaw-muscle movements (*Marcus Gunn jaw-winking phenomenon*).
6. Visual functions and refractive errors to exclude amblyopia.
7. Pupillary examination because miosis occurs in Horner's syndrome and mydriasis in the oculomotor nerve palsy.
8. Presence of telecanthus, epicanthus inversus and shortening of the lids which are often associated with severe congenital ptosis (*blepharophimosis syndrome*).
9. Photographic record that is helpful in initial evaluation and follow-up examinations.
10. Pharmacological testing to diagnose Horner's syndrome and myasthenia gravis:
    a. *Cocaine drop test*: The pupil affected by Horner's syndrome does not dilate with topical 4% cocaine because of the absence of norepinephrine. In contrast, the pupil dilates in the normal eye due to the availability of norepinephrine as a result of blockage of reuptake of norepinephrine by cocaine.
    b. *Tensilon test*: Infusion of tensilon (edrophonium chloride) results in the movement of the ptotic lid. The enzyme acetylcholinesterase breaks down acetylcholine after the muscle is stimulated thereby preventing prolonged muscle response to a nerve impulse. Tensilon temporarily blocks the action of acetylcholinesterase. However, the test has potential side effects. Therefore, other

simple approaches like *icepack test* (an application of icepack to the ptotic lids for 2 minutes inhibits acetylcholinesterase activity) and *acetylcholine receptor antibody test* (antibodies are detected in 90% of patients with systemic myasthenia gravis and 70% of patients with ocular myasthenia) have replaced the tensilon test.

In congenital bilateral partial ptosis, an attempt is always made to elevate the ptotic lids by overaction of the frontalis. The patient often tilts the head backwards or rotates the eyes downwards in an attempt to have a better vision. Some degree of ptosis may be masked by these efforts. The presence of wrinkling on the forehead indicates frontalis overaction. The upper lid crease is suggestive of some action of LPS.

Management

The management of ptosis is variable, consequent upon the cause. Ptosis due to the paralysis of the oculomotor nerve must be treated on conservative lines for 6 to 9 months. Complete paralysis of the III cranial nerve usually causes ophthalmoplegia and the elevation of the ptotic lid leads to manifest diplopia. Hence, the operation is contraindicated.

Myogenic ptosis particularly due to myasthenia gravis responds to corticosteroids, anticholinesterase agents like pyridostigmine, neostigmine or physostigmine, immunosuppressive therapy (azathioprine or cyclosporine A), plasma exchange and thymectomy.

In unilateral or bilateral congenital and mechanical ptosis, the deformity is usually corrected by an operation. Before any ptosis surgery is contemplated, the action of LPS should be carefully assessed. Depending on the levator function, the following operative techniques may be utilized.

1. When the levator action is weak, the muscle may be strengthened by its shortening either by conjunctival route (*Blaskovics' operation*) or by skin approach (*Everbusch's operation*) (video). A levator resection of 14 to 17 mm is adequate to correct a moderate degree of ptosis. A maximum resection of 23 mm of levator is advocated in a severe degree of ptosis associated with poor levator function. But the results are often poor.
   In mild degree of ptosis with good levator function, Fasanella-Servat operation (tarsoconjunctival resection) (video) gives good cosmetic result. The operative procedure is quite simple.
2. When the LPS is completely paralyzed but the superior rectus is functioning, the middle third of the tendon of superior rectus is transplanted to the upper border of the tarsal plate (*Motais' operation*).
3. The frontalis sling operation is preferred over Motais' operation. The elevating effect of the frontalis muscle is utilized in the operation. The lid is slinged to the frontalis muscle by passing 3-0 polypropylene sutures or fascia lata strips (video) subcutaneously *(Hess' operation* or *frontalis suspension)*. Bilateral Hess' operation is indicated in a patient with jaw-winking phenomenon after the disinsertion of the LPS.

TUMORS OF THE LIDS

Both benign and malignant tumors of the skin and glands of the lids may occur.

Benign Tumors

Common benign tumors of the lids are nevus, hemangioma, xanthelasma, Molluscum contagiosum and neurofibromatosis.

Nevus

Nevus or mole may affect either the skin or the conjunctiva of the lid. The nevus is composed of nevus cells arranged in an alveolar manner. Most

nevi remain stationary, but they may grow at puberty and, rarely, undergo malignant transformation.

Hemangioma

Hemangioma of the lid (Fig. 24.23) is not uncommon and occurs usually in two forms, capillary and cavernous.

The *capillary hemangioma* manifests as bright red or portwine spots composed of dilated capillaries, while the *cavernous hemangioma* consists of large, dilated and anastomosing subcutaneous venous channels which are blue in color. Sometimes, hemangioma is found in the distribution of first and second divisions of the trigeminal nerve and is associated with hemangiomata of choroid and leptomeninges and hydrophthalmos (Sturge-Weber syndrome).

Treatment

Small hemangioms may be left alone as spontaneous involution is the rule, but if they increase in size and interfere with vision, treatment is necessary. Local intralesional injection of a mixture of 40 mg of triamcinolone acetonide and 6 mg of betamethasone sodium phosphate into the hemangioma may lead to its regression. Administration of systemic corticosteroids may cause involution of the growth in some cases. Superficial radiotherapy (100-200 rads) monthly for 6 months gives encouraging results. Some cases need surgical excision using a cutting diathermy.

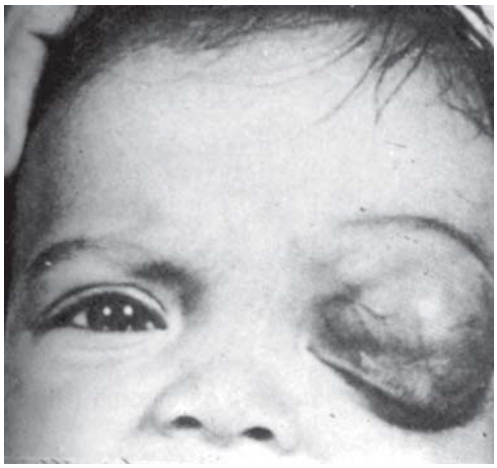

Fig. 24.23: Hemangioma of left upper lid

Papilloma

Various benign epithelial proliferations in the lid are grouped under the heading of papilloma. Squamous papilloma is the most common benign tumor of the lid. It is seen in patients older than 30 years due to exposure to ultraviolet light. It may present as a round multilobular lesion that can be sessile or pedunculated with a central vascular core. Electrocautery, chemical cauterization and CO_2 laser ablation can be used to remove squamous papilloma.

Seborrheic keratoses may be sessile or pedunculated and have a varying amount of pigmentation and hyperkeratosis. Exuberant hyperkeratosis presents as a cutaneous horn.

Xanthelasma

Xanthelasmas are often bilateral, symmetrical and appear as slightly raised yellow wrinkled plaques near the inner canthus. They probably represent lipid deposits in histiocytes in the dermis of the lid. They are frequent in elderly women and occasionally found associated with diabetes and hypercholestrolemia. The lesion may be excised or destroyed by cryotherapy.

Molluscum Contagiosum

Molluscum contagiosum is caused by a large-sized poxvirus. The lesion is characteristically small waxy and nodular with a central umblication often involving the lid margin. It may cause a toxic conjunctivitis or a mechanical pannus. The growth should be excised and cauterized by pure carbolic acid.

Neurofibromatosis

Neurofibromatosis is a generalized disease that may involve the lid and cause mechanical ptosis (Fig. 24.24). It may occasionally be associated with unilateral infantile glaucoma. Small, multiple tumors are distributed along the hypertrophied nerves. *Café au lait* spots are often present.

Malignant Tumors

Carcinoma, sarcoma and malignant melanoma are the malignant tumors of the lids.

Carcinomas of the lid are more common than the other tumors and occur most frequently in men over the age of 50 years. Carcinoma of the lid is usually of two types—basal cell carcinoma and squamous cell carcinoma .

Basal Cell Carcinoma

Basal cell carcinoma or rodent ulcer (Fig. 24.25) is the most common malignant eyelid tumor predominantly affecting the lower lid. Basal cell carcinoma is seen in young patients with a positive family history of systemic malignancy. It is locally invasive and does not spread to the regional lymph nodes.

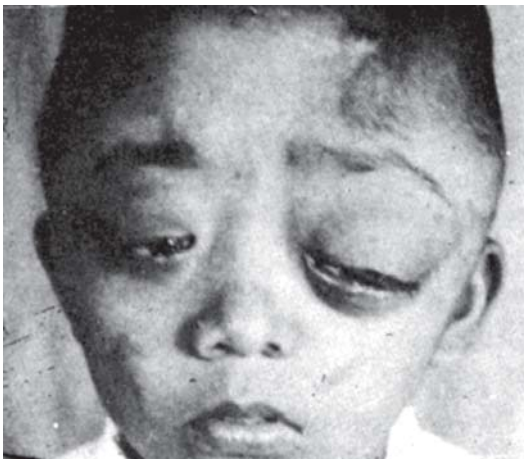

Fig. 24.24: Neurofibromatosis with proptosis and ptosis of left eye and *café au lait* spots

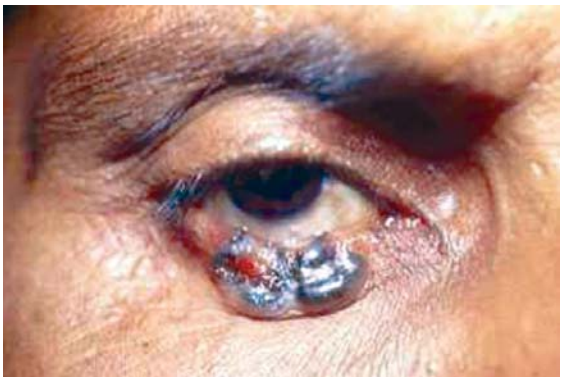

Fig. 24.25: Basal cell carcinoma of lower lid (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

The tumor starts as a small nodule which ulcerates. The ulcer has a raised nodular border and an indurated base. It spreads very slowly and eventually erodes the surrounding structures.

The less common variety of basal cell carcinoma may start as a firm and elevated lesion with undeterminate borders (*fibrosing basal cell carcinoma*). This type of carcinoma behaves more aggressively than the nodular type.

Treatment

Surgical excision is the treatment of choice for all basal cell carcinomas of the eyelid. Cryotherapy is indicated for those patients who are unable to tolerate the surgery. A high recurrence rate and depigmentation of the lid are two main disadvantages of the cryotherapy. Radiotherapy is considered as a palliative treatment. It should not be used for canthal lesions. The therapy may lead to obstruction of the lacrimal drainage system and radiation induced injury to the eyeball.

Squamous Cell Carcinoma

Squamous cell carcinoma (epithelioma) shows a predilection for the lid margin (Fig. 24.26). It usually starts as a small nodule which ulcerates. It grows slowly and painlessly. The base of the ulcer is indurated. Later, the tumor enlarges in

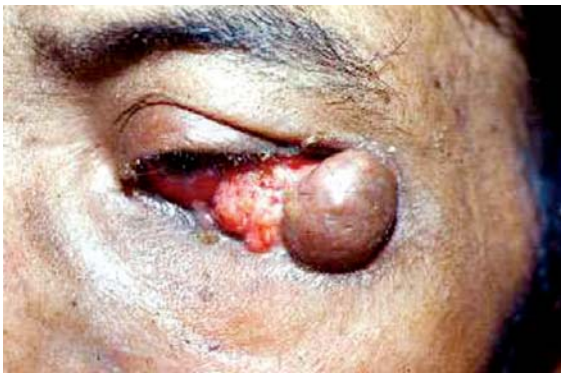

Fig. 24.26: Squamous cell carcinoma
(*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

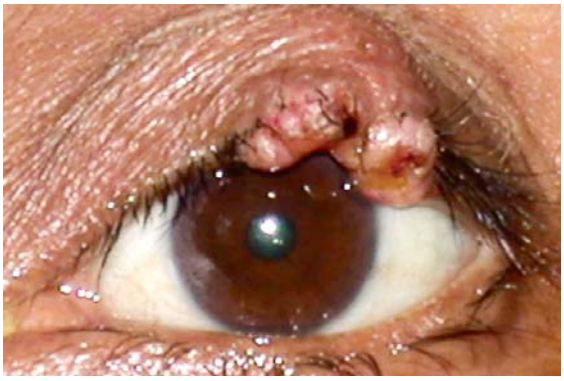

Fig. 24.27: Sebaceous gland carcinoma of upper lid
(*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

size resulting in a fungating growth. The regional lymph glands (preauricular and submandibular) are often enlarged. Metastases are common and occur through lymphatics. The growth should be excised early followed by radiotherapy.

Sebaceous Adenocarcinoma

Sebaceous gland carcinoma (Fig. 24.27) arises from the meibomian glands in the tarsal plate. It is a higly malignant tumor. A nodule in the upper eyelid, initially simulating as a chalazion, destroys the meibomian gland orifice. The tumor metastasizes to the regional lymph nodes. A full-thickness biopsy helps in the diagnosis. Wide surgical excision is needed.

Sarcoma of the lid is quite rare. Orbital lymphoma, lymphosarcoma or rhabdomyosarcoma may involve the lid by anterior extension. Most of these tumors are radiosensitive.

Malignant Melanoma

Malignant melanoma of the lid may develop *de novo* or from a pre-existing melanotic nevus. Eyelid involvement may be in the form of superficial spreading malignant melanoma, lentigo maligna melanoma (invasive) or nodular melanoma. A radical excision of the lid should be performed early.

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CHAPTER

25 Diseases of the Lacrimal Apparatus

ANATOMY

The lacrimal apparatus consists of the lacrimal gland, the accessory glands and the lacrimal passage.

Lacrimal Gland

The lacrimal gland is a tear-secreting gland comprising the superior (orbital) and the inferior (palpebral) part.

The *superior lacrimal gland* is of the size and shape of an almond situated in the lacrimal fossa at the outer part of the orbital plate of the frontal bone.

The *inferior lacrimal gland*, comprising only 1 or 2 lobules, lies just above the lateral part of the upper fornix. There are about 12 ducts collecting the secretion of the whole gland. They open upon the surface of the conjunctiva at the outer part of the upper fornix. Histologically, the lacrimal gland consists of acini and ducts and resembles the salivary gland.

The *accessory lacrimal glands of Krause* are microscopic groups of acini lying in the conjunctival mucosa between the fornix and the edge of the tarsus. They are approximately 42 in the upper fornix and 6-8 in the lower. They form a common duct to open into the respective fornix. Extirpation of the lacrimal gland does not lead to dryness of the conjunctiva as secretions from the goblet cells and Krause glands are sufficient to wet the conjunctiva.

The lacrimal gland is supplied by the lacrimal branch of ophthalmic artery.

The trigeminal, the facial and sympathetic nerves supply the lacrimal gland.

Lacrimal Passage

The lacrimal passage (Fig. 25.1) consists of lacrimal puncta, lacrimal canaliculi, lacrimal sac and nasolacrimal duct.

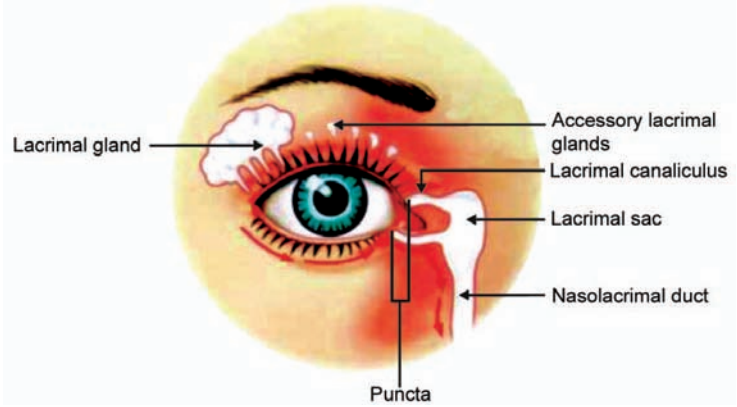

Fig. 25.1: Anatomy of lacrimal passage (*Courtesy*: Allergan India)

The *lacrimal puncta* are two openings lying near the posterior margin of each lid about 6 mm away from the inner canthus. The punctum is situated upon a slightly elevated area, the *lacrimal papilla*.

The *lacrimal canaliculus* joins the punctum to the lacrimal sac. It first passes vertically for about 1 to 2 mm and then horizontally 6 to 7 mm. The superior and the inferior canaliculus meets to form a common canaliculus which opens in the lateral wall of the lacrimal sac.

The *lacrimal sac* lies in the lacrimal fossa formed by the lacrimal bone and the frontal process of the maxilla. When distended it measures about 15 mm in length and 5 to 6 mm in breadth. The upper portion of the sac, i.e., the fundus, extends 3 to 5 mm above the medial palpebral ligament. The lower end of the sac narrows down and continues as nasolacrimal duct (NLD).

The nasolacrimal duct is about 15 mm in length and has a diameter of about 3 mm. It runs downwards, slightly outwards and backwards, and opens in the anterior part of the outer wall of inferior meatus of the nose. The course of the duct may be represented by a line joining the medial angle of the eye to the first upper molar tooth. There are folds of mucous membrane in the nasolacrimal duct. The upper end of the duct is the narrowest part.

The lacrimal sac and the nasolacrimal duct are lined by columnar epithelium, while the canaliculi by stratified squamous epithelium. The lower end of the duct may remain covered by a membrane, especially in the newborn.

DISEASES OF THE LACRIMAL GLAND

The lacrimal gland may be implicated in inflammatory, traumatic, obstructive and neoplastic lesions.

Dacryoadenitis

The inflammation of lacrimal gland is known as *dacryoadenitis*.

Types

Dacryoadenitis may occur in two forms: (i) acute dacryoadenitis, and (ii) chronic dacryoadenitis.

Acute Dacryoadenitis

Etiology

Acute dacryoadenitis is usually associated with mumps, measles or infectious mononucleosis.

Clinical Features

It is a painful condition, and manifests as swelling of the upper lid margin having a typical S-shaped curve due to the involvement of the palpebral part of the lacrimal gland. Preauricular lymphadenopathy almost always accompanies the disease.

Chronic Dacryoadenitis

Chronic dacryoadenitis is relatively more common than the acute dacryoadenitis.

Etiology

Granulomatous diseases such as tuberculosis and sarcoidosis may cause chronic dacryoadenitis, the latter perhaps is the most common cause.

Clinical Features

Chronic dacryoadenitis is characterized by painless, non-tender swelling of the lacrimal gland associated with localized edema and mild ptosis of the upper lid.

Treatment

Dacryoadenitis can be managed by hot compresses, systemic NSAIDs and appropriate local and systemic antimicrobial therapy (depending upon the cause). Noninfectious cases such as sarcoidosis need systemic corticosteroids. The lacrimal gland abscess generally requires surgical drainage.

Mikulicz's Syndrome

Mikulicz's syndrome is characterized by bilateral symmetrical enlargement of the lacrimal and salivary glands. Sarcoidosis and tuberculosis are usually associated with the syndrome. The exact cause of Mikulicz's syndrome is not known but it is believed to be an autoimmune disorder and a variant of Sjogren's syndrome.

Neoplasia of the Lacrimal Gland

The lacrimal gland neoplasms may be benign or malignant.

Benign mixed-cell tumor of the lacrimal gland (pleomorphic adenoma) is the most common epithelial tumor occurring around 50 years of age. A firm painless mass occupies the lacrimal fossa and causes proptosis. It is slow growing, nontender, and displaces the eyeball downwards and medially (Fig. 25.2). CT scan is helpful in the diagnosis of the tumor (Fig. 25.3). The mixed cell tumor presents a pleomorphic appearance on histopathology (Fig. 25.4). It needs a wide complete excision including the periorbita or the bone without disruption to avoid seeding of the tumor in adjacent tissues.

Adenoid cystic carcinoma is a rapidly growing tumor of lacrimal gland. It occurs between 40 and 60 years of age and presents as a painful swelling in lacrimal area. The posterior extension of tumor causes restricted motility, optic disk edema and choroidal folds. CT scan shows invasion of bone and calcification in the tumor. The prognosis for

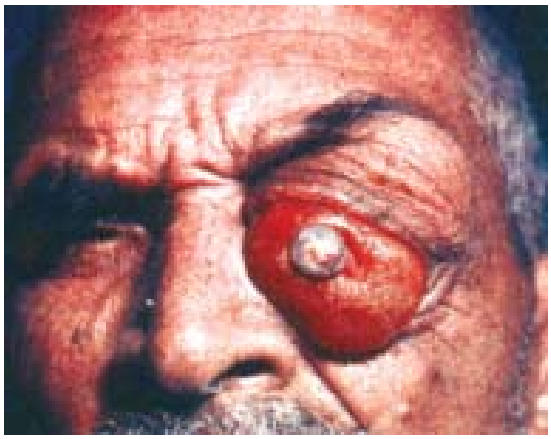

Fig. 25.2: Tumor of lacrimal gland

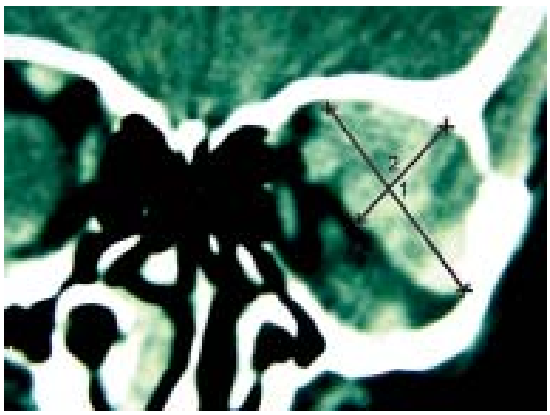

Fig. 25.3: CT of the orbit showing a mass occupying the superotemporal part of the orbit (*Courtesy*: Dr MS Bajaj, Dr RP Centre, New Delhi)

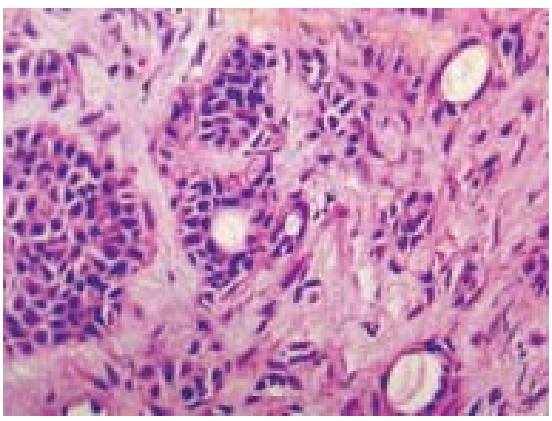

Fig. 25.4: Pleomorphic adenoma of the lacrimal gland (*Courtesy*: Dr MS Bajaj, Dr RP Centre,New Delhi)

life is poor despite orbital exenteration and radiotherapy.

PRECORNEAL TEAR FILM

The tears are a mixture of secretions from the lacrimal gland, accessory lacrimal glands, goblet cells and meibomian glands (Fig. 25.5). They are slightly alkaline having an average pH of 7.25. The rate of secretion is about 1.2 µl/minute. Tears contain globulins, bacteriostatic lysozymes, immunoglobulins, complement, glucose and electrolytes.

The tears form a thin (8 µm) layer over the cornea and the conjunctiva, it is known as *precorneal tear film* (Fig. 25.6). Blinking maintains a continuous tear film over the ocular surface. The tear film is composed of three layers.

1. The outermost *lipid layer* contains lipids and waxy esters produced by meibomian glands and glands of Zeis. The layer prevents the evaporation of aqueous part of the tear and helps in the stability of the tear film by increasing the surface tension.
2. The middle *aqueous layer* contains inorganic salts, glucose, glycoproteins, and is secreted by the lacrimal and accessory lacrimal glands. It has some buffering ability.
3. The innermost *mucous layer*, composed of hydrated mucoproteins, is secreted by goblet cells. It converts the corneal epithelium from a hydrophobic to a hydrophilic structure.

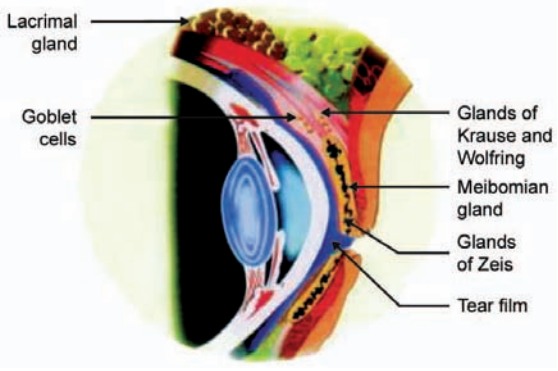

Fig. 25.5: Different types of glands contributing in formation of tear film

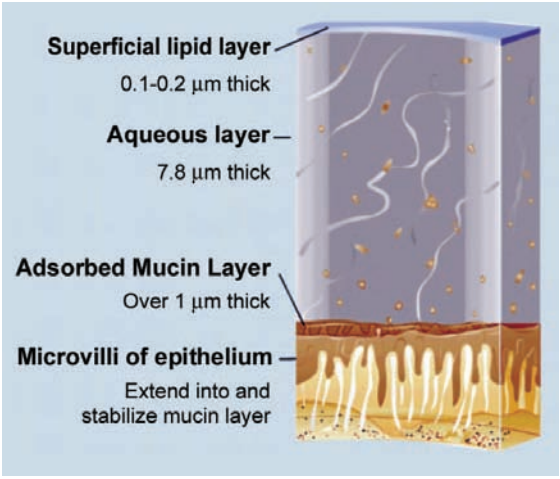

Fig. 25.6: Precorneal tear film
(*Courtesy*: Dr Vinay Agrawal, Mumbai)

Functions of Tear Film

The main functions of the tear film are to:

1. Make the cornea a smooth optical surface
2. Moisten the surface of the cornea and the conjunctiva
3. Inhibit the growth of micro-organisms by mechanical flushing and antimicrobial action of the lysozymes
4. Transport oxygen and carbon dioxide to and from the epithelial cells, and
5. Dilute and wash away the noxious stimuli.

Drainage of Tears (Lacrimal Pump)

Normally the tears are collected in the lacus lacrimalis. They are sucked into the lacrimal puncta and canaliculi partly by the capillary action and partly by the contraction of pretarsal part of orbicularis oculi muscle during the closure of the lids. The bulk of tears (70%) are drained through the lower canaliculus. The contraction of preseptal part of orbicularis oculi muscle (Horner's muscle) expands the cavity of the

lacrimal sac resulting in a negative pressure which helps to draw in the tears. The tears are then drained from the sac through the NLD into the inferior meatus due to gravitational force and relaxation of Horner's muscle during opening of eyelids.

Watering or Tearing

Watering of the eye can be divided into two broad groups: (i) hyperlacrimation, and (ii) epiphora. Ordinarily, the amount of tears produced is just sufficient to moisten the eyeball, and it is lost by evaporation. Reflex irritation often causes *lacrimation*, while obstruction in the lacrimal passage results in *epiphora*.

Hyperlacrimation

Hyperlacrimation is a condition of oversecretion of tears. It may be of 3 types:

1. Primary hyperlacrimation
2. Reflex hyperlacrimation, and
3. Central hyperlacrimation.

Primary hyperlacrimation may be caused by an irritative lesion of the lacrimal gland (inflammatory, cystic or neoplastic).

Reflex hyperlacrimation is more common and can occur due to stimulation of branches of the V cranial nerve supplying the ocular structures. It is frequently seen in conjunctivitis, corneal disorders, uveitis and glaucoma.

Central hyperlacrimation is almost invariably found in emotional states.

Epiphora

Impairment of the drainage of tears results in epiphora. Obstruction of the lacrimal passage is the most common cause of epiphora. Occasionally the lacrimal pump may not function normally due to the laxity of orbicularis oculi muscle.

DRY EYE SYNDROME

Deficiency of tears or instability of tear film causes dry eye syndrome which is a leading cause of ocular discomfort.

Etiology and Types

Dry eye syndrome (DES) occurs in different forms and may have the following causes:

1. Aqueous tear deficiency
2. Mucin deficiency
3. Lipid deficiency
4. Impaired lid function or blinking, and
5. Irregularity of the corneal surface.

The *aqueous tear deficiency* (ATD) is commonly found in keratoconjunctivitis sicca (KCS). The sicca may occur in many conditions such as Sjögren's syndrome, sarcoidosis, atrophy or hypoplasia of lacrimal gland and Riley-Day syndrome.

Sjögren's syndrome is an autoimmune disorder occurring in women of menopausal age and is characterized by dry uncomfortable eye, blepharoconjunctivitis, epithelial erosion, filamentary keratitis and fibrotic lacrimal gland. Vital staining with rose bengal reveals triangular staining of the bulbar conjunctiva on either side of the limbus, and staining of corneal filaments and mucous threads.

The *mucin (lacrimal surfactant) deficiency* results from goblet cell dysfunction. The mucin layer deficiency decreases the wettability of the ocular surface. It causes instability of the tear film and decrease in the tear film break-up time.

The important causes of mucin deficiency are: (i) hypovitaminosis A, (ii) excessive conjunctival scarring due to trachoma and membranous conjunctivitis, (iii) mucocutaneous disorders—ocular pemphigoid, erythema multiforme and Stevens-Johnson syndrome, and (iv) chemical burns and injuries.

The *lipid deficiency* can occur in the patients with chronic blepharitis and acne rosecea.

Impaired lid function and *abnormal blinking* are important causes of DES. Normal blink reflex maintains a normal tear film. Decreased blinking, incomplete closure of lids (Bell's palsy), dellen, pterygium, ectropion of the lower eyelid and neuroparalytic keratitis may adversely affect the tear film stability.

Irregularity of corneal surface (epitheliopathy) produces irregularity of the tear film. The mucous layer uniformly coats the corneal epithelium in a normal eye. However, in the disorders of the corneal epithelium, the mucous layer becomes thin and retracts in areas of epithelial irregularity.

A number of drugs like atropine, antihistaminics, hypnotics and tranquilizers, decrease the tear production.

Clinical Features

A host of symptoms like ocular discomfort, foreign body sensation, burning, blurred vision, photophobia, heaviness of lids, mucous discharge, redness and inability to open eyes in the light may occur.

The patient may present with features of nonspecific blepharoconjunctivitis. The discharge is scanty with mucous strings and debris. The bulbar conjunctiva looks dry and lusterless. In advanced cases superficial punctate keratitis, corneal mucous plaque, marginal corneal thinning, band-shaped keratopathy and corneal ulcer may develop.

Diagnostic Tests

Tear secretion, stability of tear film and morphology and density of goblet cells can be ascertained by certain diagnostic tests. They include Schirmer test I and II, tear film break-up time, conjunctival impression cytology and rose bengal staining.

Schirmer Test

Schirmer test measures the aqueous production and gives a gross idea of tear film function.

Schirmer test I is performed with the help of a 5 × 35 mm strip of Whatman filter paper in a semi-dark room. A 5 mm bent part of the strip is kept in the lower fornix at the junction of medial two-thirds and lateral one-third of the lower lid (Fig. 25.7). The patient is advised to keep his eyes open. The extent of wetting of the strip from its bent portion is noted after 5 minutes. A value between 10 and 30 mm indicates normal secretion, less than 5 mm suggests impaired secretion and greater than 30 mm hyper reflex secretion.

Basic secretion is measured after instillation of a topical anesthetic agent in the conjunctival sac, the rest of the procedure is same as described above. The amount of reflex secretion can be obtained by deducting the basic secretion from the reading of Test I.

Schirmer test II is seldom performed and it measures the reflex tear secretion. It is conducted by putting a topical anesthetic agent in the conjunctiva and irritating the nasal mucosa. The amount of moistening of the strip is recorded after 2 minutes. A reading less than 15 mm suggests failure of reflex secretion.

Tear Film Break-up Time

The mucin deficiency can be diagnosed by tear film break-up time (BUT). After instillation of

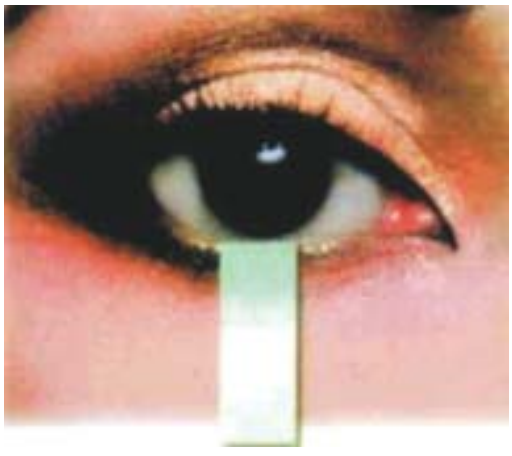

Fig. 25.7: Schirmer test

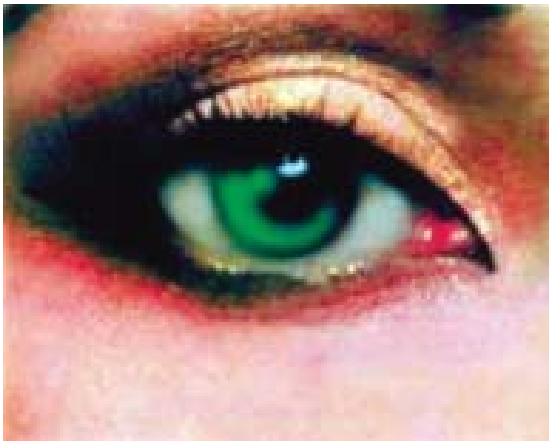

Fig. 25.8: Fluorescein staining

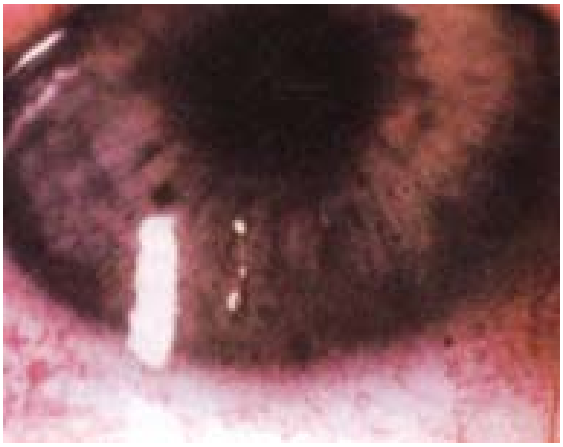

Fig. 25.10: Rose bengal staining
(*Courtesy*: Dr Vinay Agrawal, Mumbai)

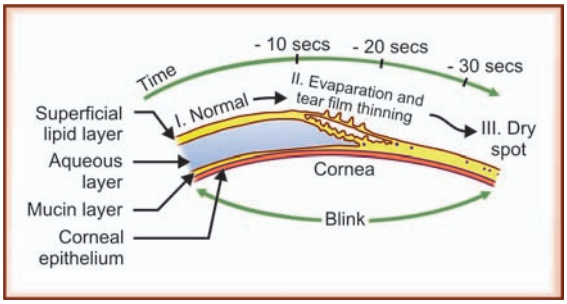

Fig. 25.9: Mechanism of tear film break-up

fluorescein the patient is asked to blink once and examined on the slit-lamp using a cobalt blue filter (Fig. 25.8). The time interval between the last blink and the appearance of first dry spot is measured with a stop watch. In the healthy eyes, BUT ranges between 15 and 35 seconds (Fig. 25.9). A time of 10 seconds or less is considered abnormal. Tonge's break-up time toposcope can be used to evaluate the image of a reflected grid. Tear breaking is seen as a distortion of an otherwise regular grid.

Conjunctival Impression Cytology

Conjunctival impression cytology (CIC) is a non-invasive technique. It can provide valuable information about the morphology and density of mucous producing goblet cells. CIC is often preferred over the conjunctival biopsy.

Rose Bengal Staining

The rose bengal staining indirectly ascertains the reduced tear secretion by detection of damaged epithelial cells. Rose bengal, a vital dye, stains the dead and dying epithelial cells. A positive test reveals a triangular staining of the nasal and the temporal bulbar conjunctiva in the exposed interpalpebral area. A punctate staining in the lower two-thirds of the cornea may occasionally be seen (Fig. 25.10). The test is useful in the diagnosis of keratoconjunctivitis sicca.

Management

The management of dry eye is not always satisfactory. Four approaches are commonly used in its management:

1. Supplementation of tears
2. Preservation of existing tears
3. Stimulation of tears, and
4. Treatment of inflammatory process.

Preservative-free tear substitutes remain the mainstay of the treatment of dry eye. Artificial tears (hypromellose and methylcellulose) are instilled frequently. Mucomimetic polymers (natural tears) are useful in both mucin and aqueous deficiency states. The slow release artificial tears pellet (lacrisert) may be inserted in the lower fornix which provides a continuous source of tears.

Puncta may be blocked in order to preserve the scanty tears.The punctum can be blocked by punctal plugs, collagen implants, argon laser punctoplasty or cauterization. Moist chamber goggles and soft contact lenses may relieve discomfort in many cases.

Some drugs such as bromhexine and eledoisin stimulate the lacrimal gland for the production of more tears.

Treatment of inflammatory process also relieves the symptoms. A chronic immune-mediated inflammatory process plays a role in the pathogenesis of dry eye. Topical instillation of corticosteroids and cyclosporine A drops significantly improve the symptoms of dry eye. Systemic tetracycline is the treatment of choice in blepharitis associated with dry eye.

The production of mucous strands can be minimized by the use of acetylcystein (10%) drops. If needed, surgery may be performed to correct the lid deformities and inadequate blinking.

DISEASES OF LACRIMAL PASSAGE

Obstruction of Lacrimal Passage

Obstruction of the lacrimal passage may occur at any level—punctum, canaliculus, lacrimal sac, and nasolacrimal duct (NLD).

Punctal obstruction is quite common due to foreign body, stenosis and ocular chemical burns.

Canalicular obstruction may be congenital or acquired. Stricture of the canaliculus develops following trauma and infection.

Lacrimal sac obstruction may be congenital or acquired. Recurrent chronic dacryocystitis causes obstruction.

Nasolacrimal duct obstruction, especially the congenital, is the most common cause of epiphora in infants due to noncanalization of the duct. Chronic dacryocystitis and involutional stenosis of NLD frequently lead to watering in old people.

Work-up

Initially it is important to exclude hypersecretion of tears, reflex hypersecretion and lacrimal pump failure in a case of epiphora.

A case of epiphora needs thorough clinical examination to identify any developmental or acquired disorders of the lacrimal drainage system.

The position of puncta and lower lid, size of the puncta, presence of a foreign body or debris in the punctal orifice and a swelling or a discharging sinus over the sac region should be identified.

A slight pressure over the lacrimal sac may lead to reflux of pus or mucus through the punctum suggesting a mucocele with intact canaliculi and punctum.

It is equally important to rule out the presence of a nasal pathology (like nasal polyp or atrophic rhinitis).

Patency of the Lacrimal Passage

The patency of the lacrimal passage can be assessed by dye tests, syringing (irrigation), dacryocystography and radionucleotide dacryocystography.

Fluorescein Dye Disappearance Test: When a 2% solution of fluorescein is instilled into the normal conjunctival sac, the dye disappears after 2 minutes. The test is of great significance in patients with unilateral watering. When there occurs a retention of dye at the interface of the lower eyelid margin and the cornea (high marginal tear strip), it suggests an obstruction of the lacrimal drainage system.

Jones Primary Dye Test (The Jones Test I): It has the same principle as that of fluorescein disappearance test. However, in this test an anesthetic soaked cotton bud is placed under the inferior

turbinate of the nose and after 5 minutes the cotton bud is removed and inspected and the results are interpreted as positive or negative. In a positive test the fluorescein is recovered from the nose, while in a negative test no dye is found on the cotton bud. The latter suggests an obstruction.

Syringing (Irrigation): After anesthetization of the conjunctival sac and dilatation of the lower punctum and canaliculus with Nettleship's punctum dilator, syringing is performed to locate the site of the obstruction in the lacrimal passage. A lacrimal canula attached to a syringe filled with normal saline is passed into the lacrimal canaliculus through the lower punctum and the sac is irrigated. If the saline passes into the nose, the passage is free of obstruction, if it passes into the nose with forced pressure on the syringe, a partial obstruction is present, and if no saline reaches the nose, an obstruction is present. In the latter situation, the saline will reflux either through the upper punctum (obstruction in the sac, at the junction of the sac and the nasolacrimal duct or in the nasolacrimal duct) or through the lower punctum (obstruction in the lower or common canaliculus).

Jones Secondary Test (The Jones Test II): When syringing allows detection of fluorescein on the cotton bud placed in the nose, the test is considered positive. The positive test indicates that epiphora is not due to obstruction of the lacrimal passage but a functional failure.

Dacryocystography: The lacrimal passage can be studied radiologically by injecting a radio-opaque dye, isophendylate, into the canaliculus followed by taking posteroanterior and lateral exposures immediately. The radio-opaque dye passes through the lacrimal passage (Fig. 25.11). The exposures are repeated after 30 minutes. The retention of the dye in the lacrimal sac after 30 minutes suggests a partial or complete obstruction of the nasolacrimal duct (Fig. 25.12).

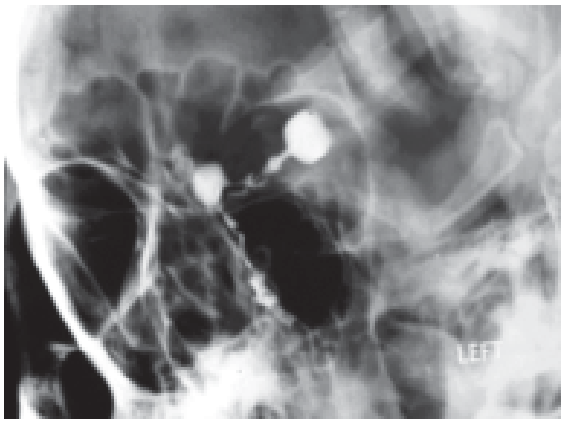

Fig. 25.11: Normal dacryocystogram showing contrast dye into the nasal cavity (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

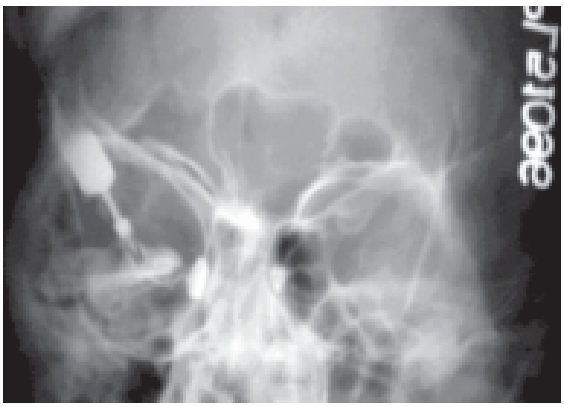

Fig. 25.12: Dacryocystogram showing pooling of dye into the lacrimal sac due to obstruction at sac- nasolacrimal duct junction (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

Radionucleotide Dacryocystography (*Lacrimal Scintillography*)*:* Radionucleotide dacryocystography is a noninvasive imaging technique to study the functional integrity of the lacrimal passage. A drop of radionucleotide tracer technitium-90m in saline is instilled into the conjunctival sac and sequential images are obtained with an Anger gamma camera. The images can be reviewed on a video screen or transferred to X-ray plates.

Eversion of the Lower Punctum

Normally, the lower punctum is not visible and the lower lid remains in apposition with the globe. Blepharitis, chronic conjunctivitis, senile laxity of the lower lid and other causes of ectropion lead to eversion of the lower punctum (Fig. 25.13) which causes epiphora. Some relief may be obtained by cauterization just behind and below the site of the punctum with a diathermy. The punctum is pulled inwards following the contraction of the fibrous tissue. Alternatively, the punctum should be slit open through its posterior border.

Occlusion of the Punctum and Canaliculus

Congenital anomalies of puncta and canaliculi such as membranous occlusion, complete absence, stenosis and reduplication, can result in watering.

The punctum and the canaliculus, particularly the lower one, may be blocked by cilium, concretion or foreign body, the blockage causes annoying epiphora.

The patency is restored by the removal of the cilium, dislodgement of concretion and sometimes by slitting the punctum and the canaliculus (*three-snip operation*).

DISEASES OF THE LACRIMAL SAC

Inflammation of the lacrimal sac is called *dacryocystitis*. It is a common disease occurring at any age. It is usually divided into two forms: congenital dacryocystitis and dacryocystitis in adult.

Congenital Dacryocystitis

Etiology

The congenital dacryocystitis or dacryocystitis of newborn (Fig. 25.14) is usually caused by membranous blockage of the lower end of the nasolacrimal duct (valve of Hasner) .

Clinical Features

The obstruction of the nasolacrimal duct is present in approximately 50% of the newborns at birth. However, watering does not occur immediately as lacrimation does not begin until 6 weeks after birth. Patency may be restored spontaneously in some cases after birth.

The condition may be bilateral and manifests as epiphora in newborn. Later, purulent discharge develops resulting in matting of the eyelashes. A gentle pressure over the lacrimal sac produces the reflux of purulent discharge from the lower punctum. There may be associated conjunctivitis.

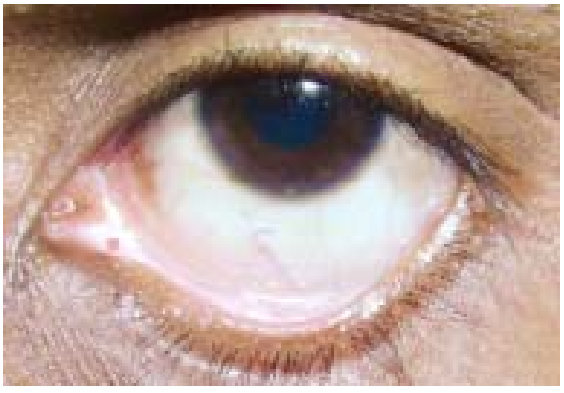

Fig. 25.13: Eversion of lower punctum

Fig. 25.14: Congenital dacryocystitis (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

Treatment

1. *Massaging* of the lacrimal sac region and frequent instillation of antibiotic drops usually cure the condition in few weeks, and the duct becomes patent. The massage increases the hydrostatic pressure and helps rupture the membranous obstruction. To obtain an effective hydrostatic pressure in the sac, the following technique is employed. The index finger is kept over the common canaliculus to prevent the regurgitation through the puncta, then it is stroked downwards firmly 10-12 times at one sitting. The procedure is repeated 4 times a day. Each massage is followed by instillation of antibiotic drops. The hydrostatic massage may cure the congenital obstruction in about 95% of cases.
2. *Probing* of the lacrimal passage is warranted in failed cases. But it should not be undertaken until the age of 6 months. Probing is usually performed under general anesthesia. It may be carried out either through the upper or the lower punctum with a Bowman's lacrimal probe. Postoperatively, steroid-antibiotic drops should be instilled 4 times a day. Repeat probing may be performed if epiphora persists even after 4 weeks. Generally, the success rate of probing is quite high, but it decreases if the procedure is done after the age of 18 months. Repeated probing may lead to the formation of stricture of the nasolacrimal duct. Untreated cases can develop mucopurulent conjunctivitis and lacrimal abscess. Turbinate infracture, intubation with silicon tube for 6 months, balloon dacryoplasty and dacryocysto-rhinostomy (DCR) surgery may be needed in recalcitrant cases.

Dacryocystitis in Adults

The dacryocystitis in adults may occur in an acute or a chronic form.

Acute Dacryocystitis

Acute dacryocystitis is an acute suppurative inflammation of the lacrimal sac.

Etiology

Acute dacryocystitis may occur due to various causes, the commonest being the complete obstruction of NLD. The chronic stasis of tears in the sac leads to secondary infection by *Streptococcus pneumoniae*, *Streptococcus pyogenes*, *Staphylococcus aureus* and *Actinomyces.* Acute dacryocystitis is usually preceded by a chronic dacryocystitis or an infective conjunctivitis.

Clinical Features

Acute dacrocystitis is characterized by severe pain and marked swelling and redness of the sac region and both the eyelids. Initially the sac region is swollen, red and tender. The erythema below the medial canthus spreads to the cheek. It is often associated with tenderness and enlargement of the regional lymph nodes.

Later the sac becomes filled with pus and its distended anterior wall ruptures to give rise to a pericystic swelling (Fig. 25.15). A lacrimal abscess develops which usually points below and to the outer side of the sac owing to the gravitation of the pus. It often bursts spontaneously on the skin surface forming a lacrimal fistula.

Constitutional symptoms such as malaise and fever are not uncommon with acute dacryocystitis.

Complications

Corneal ulcer, osteomyelitis of the lacrimal bone and orbital and facial cellulitis may develop as complications of acute dacryocystitis.

Treatment

Local hot compresses several times in a day and systemic NSAIDs relieve the pain. Infection is

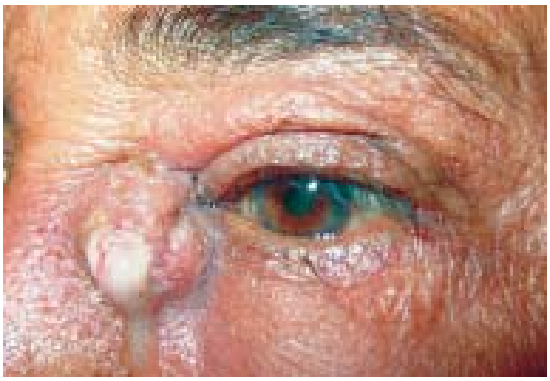

Fig. 25.15: Acute dacryocystitis (*Courtesy*: Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

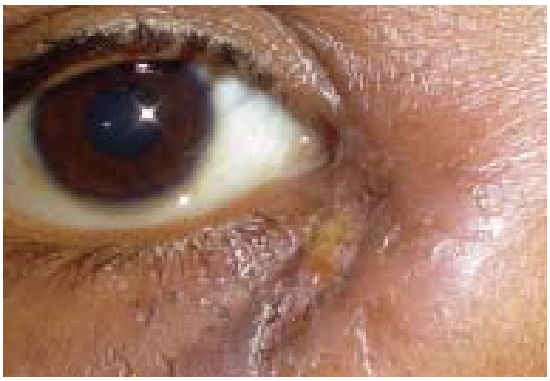

Fig. 25.16: Chronic dacryocystitis (*Courtesy:* Prof. Manoj Shukla, AMUIO, Aligarh)

controlled by topical application of antibiotic drops and ointment and systemic administration of ciprofloxacin or cephalosporins or tetracycline for seven days. In case pus point is formed, an incision is made to evacuate it. When acute inflammation subsides, the case is treated on the lines of chronic dacryocystitis. Fistulectomy with DCR operation is preferred.

Chronic Dacryocystitis

Etiology

Chronic dacryocystitis (Fig. 25.16) is more common than acute dacryocystitis and occurs following obstruction of the nasolacrimal duct due to chronic inflammation. The disease is predominantly seen in females (80%) and is usually unilateral.

Nasal pathology such as polyp, deviation of the septum, rhinitis or hypertrophied inferior turbinate are the risk factors.

Generally, the sac harbors many pathogenic organisms, viz. *Staphylococcus aureus*, *Streptococcus pneumoniae*, *Enterobacter, E. coli* and *Pseudomonas aeruginosa*. The sac may be secondarily affected from tuberculous lesion of skin, conjunctiva, nose and bones. In tertiary syphilis lacrimal sac affections are common. Mycotic dacryocystitis may occur occasionally due to *Rhinosporidium seeberi*.

Clinical Features

Persistent discharge from the eye is the main symptom of the disease. In low grade infection, a round, nontender, cystic swelling appears in the region of the sac which is known as *mucocele.* On application of pressure, a mucoid fluid regurgitates into the conjunctival *cul-de-sac* through the lower punctum. Occasionally, the fluid passes into the nose through a partially obstructed nasolacrimal duct.

Low grade repeated infections for a prolonged period of time result in a small fibrotic sac which is often associated with persistent watering and discharge. The presence of a lacrimal fistula discharging mucopus indicates a past acute suppurative dacryocystitis. Acute inflammation may supervene over chronic dacryocystitis resulting in an acute on chronic dacryocystitis.

Chronic dacryocystitis causes chronic conjunctivitis and *vice-versa*. If an intraocular operation is performed in the presence of an occult lacrimal infection, there is a risk of development of postoperative infection.

In rhinosporidiosis the sac is filled with creamy pus and its walls lined by polypoid granulation tissue containing thousands of endospores. It may be associated with rhinosporidiosis of the nose.

Complications

Chronic dacryocystitis is by far the most important contributory factor for the development of hypopyon corneal ulcer and panophthalmitis.

Treatment

The management of chronic dacryocystitis is essentially surgical. Repeated syringing of the nasolacrimal duct seldom relieves the condition.

The lacrimal drainage may be reestablished by dacryocystorhinostomy operation. Mucocele is considered to be an ideal indication for DCR surgery.

A modified dacryocystorhinostomy (with or without silastic tubing) may be needed for a failed dacryocystorhinostomy case or in patient with a fibrotic sac. If both the canaliculi are blocked with or without the nasolacrimal duct obstruction, a conjunctivodacryocystorhinostomy surgery is helpful.

Tumors of the Lacrimal Sac

Tumors of the lacrimal sac are rare. Squamous cell papilloma and transitional cell carcinoma are common tumors of the sac. The treatment includes exenteration and radiotherapy.

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CHAPTER

26 Diseases of the Orbit

ANATOMY

The eye lies in a bony cavity located on either side of the root of the nose called *orbit*. Orbit is formed by seven bones: frontal, maxilla, zygomatic, sphenoid, palatine, ethmoid and lacrimal (Fig. 26.1). Each orbit is of nearly pyramid shape and has four walls converging posteriorly. The medial walls are parallel to each other and separated by nasal cavities. The two lateral walls when extended backwards make an angle of 90° (Fig. 26.2).

Walls of the Orbit

The *medial wall* of the orbit is formed by ethmoid, lacrimal, maxillary and sphenoid bones. The thinnest wall of the orbit is lamina papyracea which covers the ethmoid sinus. This bone can be involved in blow-out fracture of the orbit.

The *lateral wall* of the orbit is formed by the zygomatic bone and greater wing of sphenoid.

The *roof* of the orbit is nearly triangular and is formed by the orbital plate of the frontal bone. The optic foramen lies at the apex of the roof.

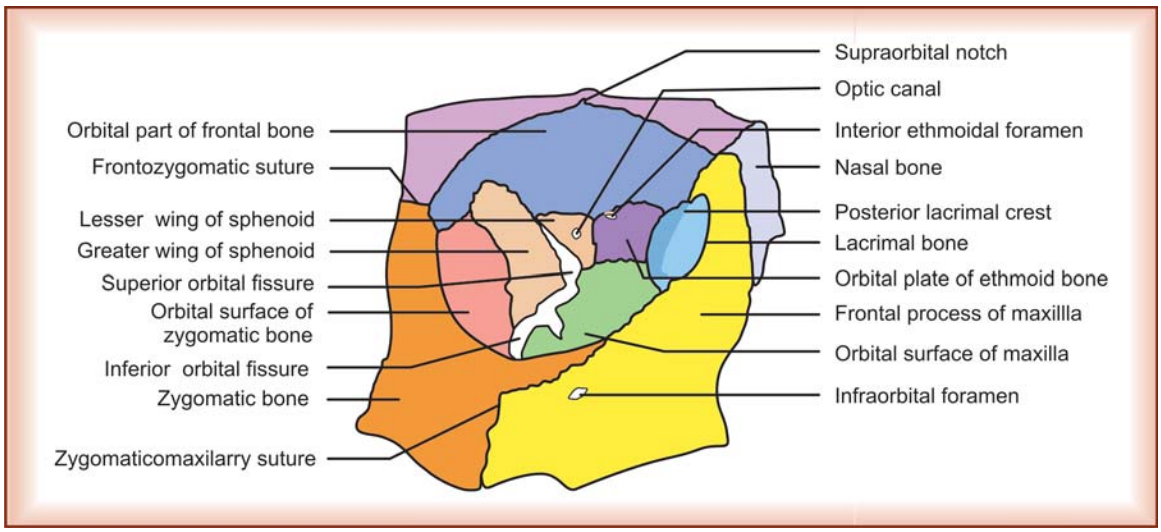


Fig. 26.1: Bones of orbit

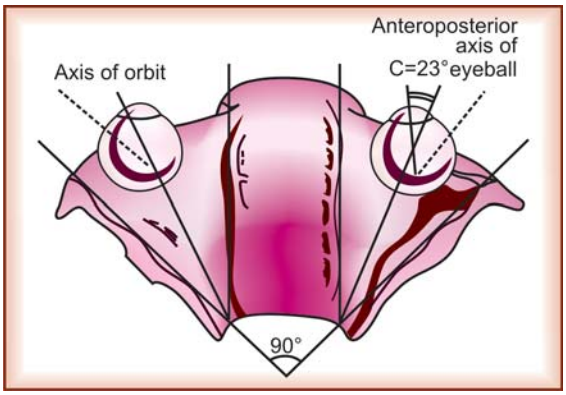


Fig. 26.2: Angles formed by orbital walls and axes of orbit

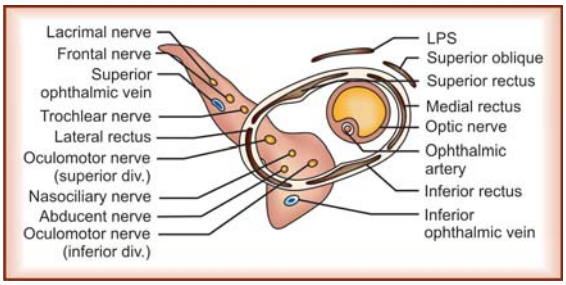


Fig. 26.3: Superior orbital fissure

The *floor* of the orbit is formed by the orbital plate of the maxilla. It is weak and often involved in the blow-out fracture of the orbit. The floor is traversed by the infraorbital groove.

Apertures of the Orbit

The orbital walls are perforated by a number of apertures, the important ones are described below.

Superior orbital fissure lies between the greater and lesser wings of sphenoid. The fissure transmits III, IV, ophthalmic division of V and VI cranial nerves as well as sympathetic fibers from cavernous plexus. The superior ophthalmic vein passes through the fissure and drains into the cavernous sinus (Fig. 26.3).

Inferior orbital fissure lies between the lateral wall and the floor of the orbit. It transmits the infraorbital nerve, zygomatic nerve, branch from the pterygopalatine ganglion and pterygoid venous plexus.

Optic canal measures approximately 10 mm and lies within the lesser wing of sphenoid. The optic nerve, ophthalmic artery and sympathetics pass through the canal. The orbital end of the optic canal is called *optic foramen* which measures about 6.5 mm in diameter.

The average volume of an adult orbit is approximately 30 ml. The eyeball occupies one-fifth of the space and rest of the orbital cavity is filled by nerves, extraocular muscles, lacrimal gland, lacrimal sac, ciliary ganglion, ophthalmic artery and vein and their branches, orbital fat and fascia.

The Orbital Fascia

The orbital fascia lines various intraorbital structures. It covers the orbital bones, and in the anterior part of the orbit forms a membrane or an intermuscular septum. The extraocular muscles do not perforate this membrane but invaginate it and the fascia is being reflected from their surface. The fascia which covers the eyeball is known as *fascia bulbi* or *Tenon's capsule*. The condensation of the fascia in the lower part of the orbit, forms a hammock on which the eyeball rests, is called the *suspensory ligament of Lockwood*.

Blood Supply

The orbit is mainly supplied by the ophthalmic artery. It is drained by the superior and inferior ophthalmic veins into the cavernous sinus, through angular vein into the facial venous system and through the inferior ophthalmic vein into the pterygoid venous plexus. There are no lymphatics in the orbit.

Surgical Spaces of Orbit

There are following 4 self-contained spaces in the orbit.

1. *Subperiosteal space* is a potential space that lies between the bones of the orbital walls and the periorbita.
2. *Peripheral space* lies between the periorbita and the extraocular muscles joined by the fascial membrane.
3. *Central space* is a cone-shaped retrobulbar space enclosed by 4 rectus muscles and their intermuscular septa.
4. *Tenon's space* is a space around the eyeball which lies between the sclera and Tenon's capsule.

DISEASES OF THE ORBIT

The diseases of the orbit, depending on the etiology, can be classified under the following heads:

1. Congenital or developmental anomalies
2. Inflammatory diseases
3. Graves ophthalmopathy
4. Orbital neoplasms, and
5. Orbital trauma.

   The involvement of orbit in ocular trauma is described in the chapter on *Injury to the Eye*.

CONGENITAL AND DEVELOPMENTAL ANOMALIES OF THE ORBIT

The developmental anomalies of the orbit are usually associated with craniofacial malformations. The craniofacial malformation can induce changes in the size, shape or position of the orbital bones and soft tissues. Relatively common developmental orbital anomalies are described below.

1. *Craniostenosis*: It is caused by premature fusion of the cranial sutures. The clinical features of craniostenosis include bilateral proptosis associated with hypertelorism(increased separation of bony orbits) and apparent divergent strabismus, papilledema and optic atrophy. The mechanical pressure on the optic nerve can be relieved by surgical decompression.
2. *Oxycephaly*: It is caused by premature fusion of the coronal sutures. The orbit becomes small, flat and elevated.
3. *Craniofacial dysostosis*: It is caused by the fusion of coronal and sagittal sutures and characterized by small orbit, proptosis, hypertelorism and skeletal deformities. Examples of craniofacial dysostosis are Crouzon syndrome and Apert anomaly.
4. *Craniofacial clefting*: A craniofacial cleft occurs when the normal development is arrested. Example of the clefting syndrome that affects the orbit and lids is mandibulofacial dysostosis *(Treacher Collins syndrome)*. It is a developmental anomaly of the first brachial arch. It is characterized by orbital deformities, antimongoloid obliquity of the palpebral fissures, coloboma of the lower eyelid, low-set ears and hypoplasia of the mandible.
5. *Meningoencephalocele*: Bones of the skull and orbit may have congenital clefts through which intracranial contents may herniate resulting in meningocele or meningoencephalocele. Meningoencephalocele is often present near the medial canthus and increases in size on crying or straining. The eyeball is usually displaced outward and downward. It may cause pulsating exophthalmos.

INFLAMMATORY DISEASES OF THE ORBIT

Inflammation of the orbit may be preseptal, or postseptal and include preseptal orbital cellulitis, periostitis, orbital cellulitis, cavernous sinus thrombosis and idiopathic orbital inflammation (pseudotumor).

Preseptal Orbital Cellulitis

In preseptal cellulitis the infection is confined to lids and periorbital structures anterior to the orbital septum (Fig. 26.4).

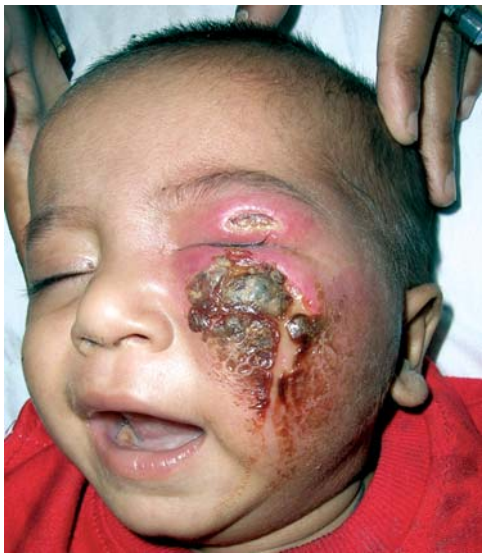


Fig. 26.4: Preseptal orbital cellulitis

Etiology

Preseptal cellulitis occurs due to penetrating trauma or secondary infection from the neighboring skin. Sinusitis is the most common cause in children. *Staphylococcus aureus* is most frequently isolated from preseptal cellulitis.

Clinical Features

The condition is often unilateral and marked by edema of the lids and periorbital swelling. Unlike orbital cellulitis, proptosis is absent and ocular movements are not restricted. CT scan of the orbit may help in the diagnosis.

Treatment

Oral or intravenous antibiotic should be administered for the control of infection. A localized abscess needs surgical drainage.

Periostitis

Periostitis usually affects the orbital margin.

Etiology

Periostitis is caused by tuberculosis, syphilis and trauma.

Clinical Features

Marginal periostitis presents a painful swelling intimately connected with the underlying bone. It tends to form an orbital abscess or progresses to orbital cellulitis. Tuberculous lesion usually results in a fistula formation. Periostitis of the deeper part of the orbit gives less defined symptoms and signs and mimics orbital cellulitis. It may cause the *orbital apex syndrome* which is characterized by ocular motor palsies, trigeminal neuralgia and anesthesia, and amaurosis owing to the involvement of the optic nerve.

Treatment

An exploratory orbitotomy may be needed for the evacuation of the pus. If untreated, the disease may extend into the cranial cavity and may cause meningitis or cerebral abscess. Prompt antibiotic therapy is necessary to control the infection.

Orbital Cellulitis

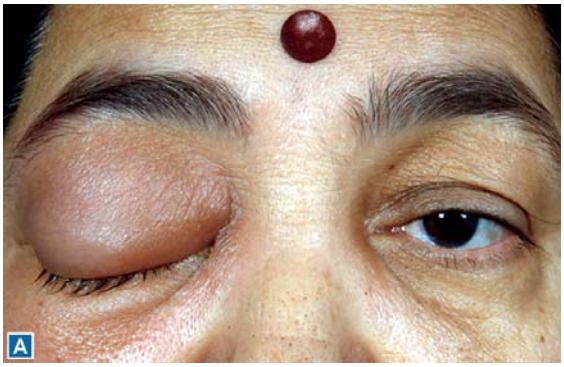
Orbital cellulitis (Fig. 26.5A) is a suppurative inflammation of the soft tissues of the orbit.

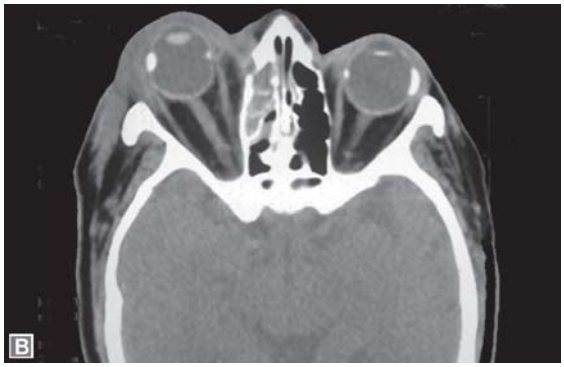
Etiology

Orbital cellulitis is usually caused by an extension of infection from the neighbouring structures particularly paranasal sinuses (90%) and teeth. It may also develop following trauma especially with a retained foreign body, septic operation on the eyeball, and facial erysipelas. *Streptococcus pyogenes, Streptococcus pneumoniae, Staphylococcus aureus, Pseudomonas aeruginosa*, *Aspergillus* and *Mucor* are the common organisms which cause orbital cellulitis.

Clinical Features

The disease is marked by severe pain, and diplopia owing to impaired ocular movements. Vision may be diminished due to retrobulbar neuritis. Swelling and redness of the eyelids, chemosis of the





Figs 26.5A and B: (A) Orbital cellulitis: marked swelling of right lid with ptosis and proptosis, (B) CT scan shows soft tissue swelling of right orbit (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

conjunctiva and varying degrees of exophthalmos are classical features of orbital cellulitis. Restriction of ocular motility and pain on movement of globe are often present. Abnormal pupillary reactions suggest orbital apex involvement.

Some cases may develop abscess formation. The pus may be localized in one of the orbital spaces and points near the orbital margin. It causes displacement of the eyeball. Ophthalmoscopy reveals engorgement of the veins and evidence of optic neuritis. Fever, headache and occasional cerebral symptoms may occur. CT scan is invaluable in the diagnosis of orbital cellulitis (Fig. 26.5B).

Complications

In neglected cases panophthalmitis, thrombosis of the cavernous sinus, purulent meningitis and cerebral abscess may supervene.

Treatment

Majority of the cases respond well to large doses of intravenous broad-spectrum antibiotics if instituted early. Subsequently the antibiotic should be tailored for specifically identified pathogens identified on cultures. The intravenous antibiotic therapy is usually continued for 10-14 days followed by oral antibiotics for an additional 14-21 days. Hot compresses are also useful. If the condition does not improve within 3 days, and CT scan shows completely opacified sinuses and presence of drainable fluid, surgical drainage must be considered.

Fungal Orbital Cellulitis

Etiology

Mucor and Aspergillus can invade the orbit. Acute mucormycosis and aspergillosis cause violent sinus infection with secondary invasion of the orbit. These fungi can also invade blood vessels and cause thrombotic vasculitis. Diabetes, malignancy and immunosuppressive therapy are known risk factors.

Clinical Features

Pain and impairment of vision are often associated with proptosis. Occasionally signs of orbital apex syndrome may be present. Nasal and palatal necrosis usually co-exist with fungal orbital cellulitis, hence the diagnosis can be confirmed by biopsy of the necrotic tissue.

Treatment

It comprises management of systemic disability, excision of the nasal mass and administration of amphotericin B (5 mg/kg, IV).

Parasitic Diseases of the Orbit

Parasitic infestations of the orbit are not uncommon especially in tropical countries. Eyelids and orbits are involved in trichinosis, cysticercosis and echinococcosis.

Trichinosis

Etiology

Trichinosis is caused by a nematode, *Trichinella spiralis*. Human beings are infected due to the consumption raw pork containing Trichinella larvae.

Clinical Features

Headache, vomiting and diarrhea are early symptoms, later a typhoid-like syndrome may develop. The larvae may induce inflammation of the eyelid, the conjunctiva and the extraocular muscle. Larval encystment in the extraocular muscles and myositis are common. Rupture of the cyst may cause severe inflammatory reaction.

Treatment

There is no specific therapy for trichinosis. However, topical and systemic corticosteroids, and oral albendazole 25 mg/kg/day or mebendazole 200-400 mg 3 times a day for 10 days may provide relief.

Cysticercosis

Etiology

Cysticercosis is caused by pork tapeworm, *Taenia solium*. The tapeworm lives in the small intestine and its head or scolex is attached to the intestinal wall. The larvae of the worm can reach the lymphatic and the blood circulation through autoinfection by ano-rectal route. Larval form of *Taenia solium* is called *cysticercus cellulosae*. It causes cysts formation in the extraocular muscles, under the conjunctiva, and in the anterior chamber, vitreous and orbit. Multiple cysts my be found in the brain.

Clinical Features

The orbital cysticercosis presents with proptosis and often mimics orbital pseudotumor. Other clinical features include restricted ocular motility, ptosis and recurrent inflammation. Ultrasonography and CT scan show focal thickening of the involved extraocular muscle, the cyst and the scolex.

Treatment

Removal of cyst by orbitotomy is the treatment of choice. Albendazole 15 mg/kg/day is given in two divided doses for 8 to 30 days. Praziquantel 50 mg/kg/day can be administered 3 times a day for 15 to 30 days. Systemic corticosteroids are given with antihelminthics to control the inflammatory reaction.

Echinococcosis

Etiology

Echinococcosis is caused by the larval form of *Echinococcus granulosus* which lives in the intestine of dogs and cats. The cyst formed by the tapeworm is called *hydatid cyst* or *echinococcal cyst*, often found in liver and lungs.

Clinical Features

The larvae of echinococcus may invade the orbit and cause proptosis and signs of space-occupying lesion in the orbit. CT scan, ultrasonography and ELISA for *Echinoccocus* antibodies help in confirming the diagnosis.

Treatment

The cyst must be removed by excision. Oral albendazole 15 mg/kg/day for 4-12 weeks or

mebendazole 50 mg/kg/day for 4 weeks should be administed in divided doses.

Diseases of Paranasal Sinuses involving Orbit

Orbit is often involved in the diseases of the paranasal sinuses.

Mucocele of the frontal sinus causes proptosis and downward displacement of the eyeball associated with edema of the upper lid.

Ethmoidal sinusitis or *polyp* displaces the eyeball laterally and cases diplopia and chemosis.

Distension or *malignancy of the maxillary sinus* causes buldging and displacement of the globe upwards. The fracture of the floor of the orbit following a blunt trauma causes classical features.

Both inflammatory and neoplastic lesions of the paranasal sinuses can lead to erosion of the bony wall of orbit.

Cavernous Sinus Thrombosis

Cavernous sinus thrombosis (CST) is not an orbital disease in strict sense.

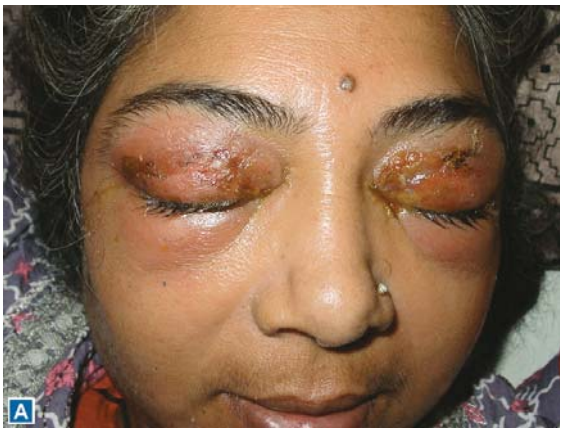
Etiology

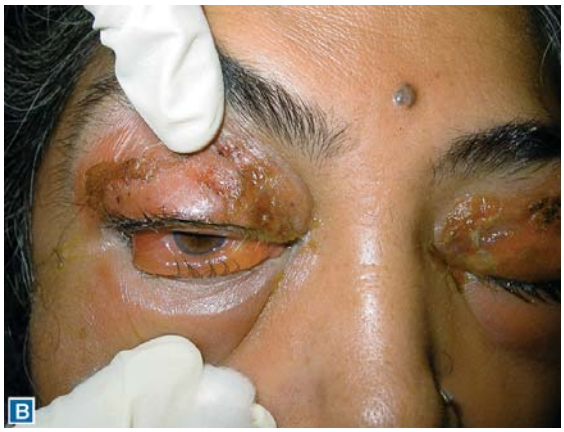
The thrombosis of the cavernous sinus may occur due to the spread of infection from orbital cellulitis, otitis, facial furuncles and erysipelas. *Staphylococcus aureus* accounts for about 70% of all infections. *Streptococcus pneumoniae* and *Aspergillus* are also implicated in causation of CST.

Clinical Features

The signs and symptoms of cavernous sinus thrombosis are almost the same as that of orbital cellulitis. Differentiation of cavernous sinus thrombosis from orbital cellulitis is difficult in the initial stages. However, the presence of edema in the mastoid region owing to the thrombosis of emissary veins, and transfer of the symptoms to the other eye (50% of cases) in the form of paralysis of lateral rectus muscle are of great diagnostic importance. Thrombosis of cavernous sinus is often accompanied by cerebral symptoms, vomiting and rigors.

Bilateral proptosis with edema of the eyelids (Figs 26.6A and B), ophthalmoplegia and corneal anesthesia are common. The paresis of the third, fourth, sixth, and ophthalmic division of the fifth





Figs 26.6A and B: Bilateral cavernous sinus thrombosis: (A) Proptosis and severe edema of eyelids, (B) Chemosis of conjunctiva and restricted opening of lid (*Courtesy:* Dr AK Grover, Sir Ganga Ram Hospital, New Delhi)

cranial nerves, dilated and fixed pupil, impaired visual acuity owing to optic nerve involvement are other features of the disease.

Diagnosis

CST is a clinical diagnosis. CT scan and MRI with contrast may be employed to confirm the diagnosis of CST and differentiate it from orbital cellulitis.

Treatment

The disease is preventable by prophylactic chemotherapy and avoidance of manipulation or squeezing of pyogenic boils over the face and nose.

Massive doses of broad-spectrum antibiotics, preferentially by intravenous route, for 3-4 weeks together with anticoagulants may control the infection and bring about the resolution. Systemic corticosteroids may be instituted under antibiotic cover to reduce inflammation and edema.

Idiopathic Orbital Inflammation or Pseudotumor of the Orbit

The idiopathic orbital inflammation, previously referred to as *pseudotumor of the orbit*, is a non-neoplastic orbital lesion characterized by a pleomorphic cellular response associated with fibrovascular reaction.

Etiology

Clinically the idiopathic orbital inflammation may present as dacryoadenitis, myositis, sclerotenonitis and Tolosa-Hunt syndrome. Inflammation of the extraocular muscles of the orbit is termed as *orbital myositis*. Besides pseudotumor, myositis is also seen in thyroid ophthalmopathy, sarcoidosis, cysticercosis and lymphoma. Chronic sclerosing-form of myositis with increased fibrosis and less inflammation occurs in *sclerotenonitis*. *Tolosa-Hunt syndrome* is caused by a nonspecific inflammation within the superior orbital fissure or cavernous sinus.

Clinical Features

Pain associated with ocular movements is highly indicative of idiopathic orbital inflammation. Visual acuity is impaired in posterior scleral involvement. Unilateral headache and severe orbital pain associated with ophthalmoplegia are characteristics of Tolosa-Hunt syndrome.

Diagnosis

Besides clinical presentation, orbital imaging (CT scan, MRI and ultrasonography) may confirm the diagnosis. The idiopathic sclerosing inflammation of the orbit may present a diagnostic dilemma since it has minimal inflammatory signs. Such lesions need biopsy for the confirmation of the diagnosis.

Both Graves ophthalmopathy and myositis cause thickening of the extraocular muscles. The former causes thickening of the muscle belly while in the latter the entire muscle is thickened along with its tendon.

Histopathologically, idiopathic orbital inflammation presents cellular infiltrates mostly consisting of lymphocytes, plasma cells and eosinophils associated with varying amount of fibrosis. An idiopathic granuloma at the apex of the orbit is found in Tolosa-Hunt syndrome.

Treatment

All cases of idiopathic orbital inflammation should be treated with high doses of systemic corticosteroids (adult dose 60-80 mg of prednisolone). Corticosteroids should be tapered more slowly over a period of several months. Bilateral cases require more prolonged therapy. Orbital irradiation (13000 cGys) and immunosuppressants (cyclophosphamide 200 mg/day) may be useful in controlling the idiopathic sclerosing inflammation of the orbit.

GRAVES OPHTHALMOPATHY

Graves ophthalmopathy is also known as *thyroid ophthalmopathy, thyroid orbitopathy* or *thyrotoxic exophthalmos.* The disease is characterized by eyelid retraction, exophthalmos (Fig. 26.7), restrictive extraocular myopathy and optic neuropathy associated with systemic features of Graves disease.

Etiology

The etiology of Graves ophthalmopathy is not known. However, it is considered as an autoimmune inflammatory disease.

Graves ophthalmopathy affects women nearly six times more frequently than men. Severity of ophthalmopathy does not parallel with the levels of T3 (tri-iodothyronine) or T4 (tetra-iodothyronine) in the serum but is closely related to the level of thyroid stimulating hormone receptor antibodies (anti-TSHR). Graves disease is basically caused by anti-TSHR and the ophthalmopathy relates to an autoimmune reaction directed towards the orbital fibroblasts.

Pathogenesis

Exophthalmos in Graves ophthalmopathy results from a discrepancy between the volume of orbit and increased volume of swollen retrobulbar tissue. There are two important interrelated reactions in the pathogenesis of Graves ophthalmopathy: (i) a cellular response in the orbit occurs through fibroblasts and preadipocyte fibroblasts, and (ii) an immunological response is mediated through TSH receptor protein expressed on orbital fibroblasts and extraocular muscles. Lymphocytic infiltration of the orbital tissue causes release of cytokines. Fibroblasts are extremely sensitive to stimulation by cytokines and immunoglobulins released during the course of an immune reaction. Stimulation of fibroblasts results in production of hyaluronic acid, a glycosaminoglycan, which increases the osmotic load and passive swelling of extraocular muscles and orbital fat.

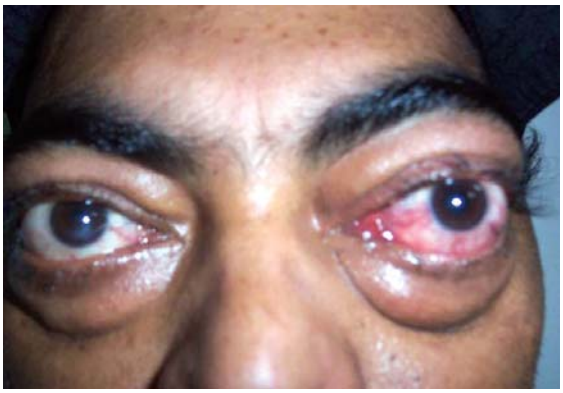


Fig. 26.7: Graves ophthalmopathy
(*Courtesy*: Dr MS Bajaj, Dr RP Centre, New Delhi)

Clinical Features

Graves ophthalmopathy is predominantly associated with Graves hyperthyroidism (90%). However, it may occur only in 1% cases of primary hypothyroidism and 6% cases with a normal functioning thyroid (euthyroid). The systemic features of the disease include tachycardia, tremors and a raised basal metabolic rate.

The most common ocular symptom of thyroid ophthalmopathy is ocular pain or discomfort which may be associated with dry eyes. Diplopia, lacrimation, photophobia and blurred vision are other symptoms of the disease.

Unilateral or bilateral eyelid retraction is the most common feature of Graves ophthalmopathy seen in more than 90% of patients. Other clinical features include unilateral or bilateral exophthalmos (Fig. 26.7), convergence deficiency (Möbius sign) restrictive extraocular myopathy and optic nerve dysfunction. Chemosis, conjunctival erythema over the insertion of medial and lateral rectus muscles, fullness of eye and superior limbic keratoconjunctivitis may be found.

Table 26.1: Characteristic eyelid signs in Graves ophthalmopathy

Clinical Feature	*Sign*	*Explanation*
Retraction of the upper eyelid	Dalrymple	Over active Müller's muscle
When eye is depressed, the upper lid lags behind	van Graefe	Over action of Müller's muscle
Eversion of the upper eyelid is difficult	Gifford	Eversion becomes difficult due to lid edema
Frequency of blinking decreases and closure becomes incomplete	Stellwag	May be due to lid edema and staring look
On looking up, the upper lid tends to move faster than the globe	Kocher	Over action of Müller's muscle
On gentle closure or taping, trembling of eyelids may be evident	Rosenbach	Sympathetic over activity
Fullness of eyelids	Enroth	Edema of lids

Various eyelid signs found in Graves disease are listed in Table 26.1.

American Thyroid Association (ATA) has classified the severity of thyroid ophthalmopathy into six grades (Table 26.2) irrespective of hormonal status. The grades can be memorized by acronym NO SPECS.

Table 26.2: ATA grading of thyroid ophthalmopathy

*Grade**	*Symptoms and signs*
0	**N**o symptom and sign
1	**O**nly signs of lid retraction and mild exophthalmopathy
2	**S**oft tissue involvement associated with lid retraction, lid lag, chemosis and exophthalmos
3	**P**roptosis is marked (23-28 mm)
4	**E**xtraocular muscle involvement
5	**C**orneal involvement
6	**S**ight loss due to optic neuropathy

*Grades 1 and 2 are considered as early and grades 3 to 6 as late ophthalmopathy

Diagnosis

The diagnosis of Graves ophthalmopathy is generally based on the following examinations and investigations.

1. Classical signs of the disease including eyelid retraction, proptosis, restrictive extraocular myopathy and optic nerve dysfunction.
2. Evidence of thyroid dysfunction or abnormal regulation.
3. Demonstration of enlargement of extraocular muscles on CT scan (Fig. 26.8), MRI or ultrasonography.
4. Testing for circulating thyroid-stimulating hormone receptor antibodies.

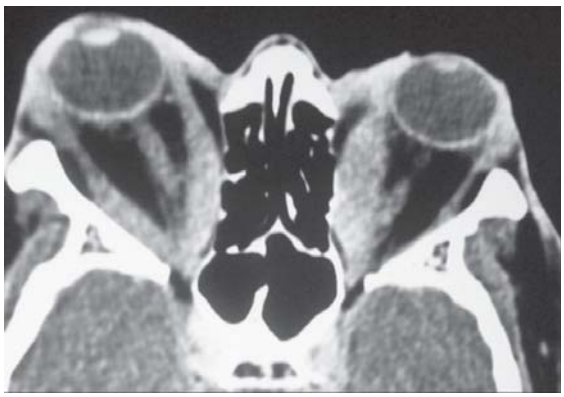


Fig. 26.8: CT scan of orbit showing marked enlargement of extraocular muscles (*Courtesy*: Dr MS Bajaj, Dr RP Centre, New Delhi)

Treatment

Graves ophthalmopathy is a self-limiting disease but it may undergo exacerbations and remissions.

Smoking has been shown to increase the ophthalmopathy. Most cases of Graves ophthalmopathy need supportive care such as ocular lubricants and tear substitutes. The management includes:

1. Correction of thyroid function abnormality with thyroxine and oral radioactive iodine (I-131) therapy.
2. Systemic corticosteroids to decrease the orbital congestion and inflammation. The pretreatment and posttreatment low-dose corticosteroids help restoration of the euthyroid state and improvement in ocular condition.
3. Surgical intervention includes:
   a. Orbital decompression to relieve the visual loss from compressive optic neuropathy
   b. Strabismus surgery to manage intractable diplopia.
   c. Correction of eyelid retraction.
4. The therapeutic value of orbital radiotherapy is questionable. It is contraindicated in diabetic patients because it can exacerbate diabetic retinopathy.

PROPTOSIS

The protrusion of the eyeball is known as *proptosis* or *exophthalmos* (Fig. 26.9). The eyeball is kept in position in the orbit by its fascial attachments and extraocular muscles. Normally, the apex of the cornea does not protrude beyond the plane of upper and lower margins of the orbit. This can be verified by putting a scale vertically on the middle of upper and lower margins of the orbit over the closed lids. The position of two eyeballs is almost always symmetrical. The protrusion of the eyeball can be accurately measured by an instrument called *exophthalmometer*.

Classification of Proptosis

Proptosis may be classified on the basis of onset, location, laterality and etiology.

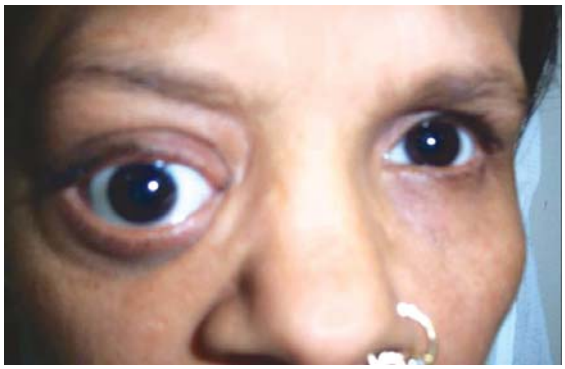


Fig. 26:9: Proptosis
(*Courtesy*: Dr MS Bajaj, Dr RP Centre, New Delhi)

Table 26.3: Common causes of proptosis

Unilateral	*Bilateral*
1. Orbital cellulitis	1. Endocrine disorder
2. Retrobulbar hemorrhage	2. Developmental shallowing of orbit
3. Orbital tumors	Oxycephaly
Glioma	Apert's anomaly
Meningioma	Crouzon's disease
Hemangioma	3. Late cavernous sinus thrombosis
Adenoma of lacrimal gland	4. Symmetrical orbital tumors
4. Orbital cysts	Lymphoma
Dermoid	Lymphosarcoma
Parasitic	Leukemic deposits
5. Early cavernous sinus thrombosis	5. Secondaries from neuroblastoma
6. Dacryoadenitis	6. Emphysema of accessory sinus
7. Orbital varix or aneurysm	
8. Pseudotumor of orbit	
9. Frontal sinus mucocele	

1. *Onset*: Acute and insidious
2. *Location*: Axial and nonaxial
3. *Laterality*: Unilateral and bilateral
4. Etiology.

Common causes of proptosis are listed in Table 26.3.

Unilateral proptosis (Fig. 26.10) may occur in orbital cellulitis, thrombosis of the orbital vein associated with or without cavernous sinus thrombosis, orbital abscess, dacryoadenitis, retrobulbar hemorrhage, intraocular tumors, orbital tumors, congenital dermoid or parasitic cysts.

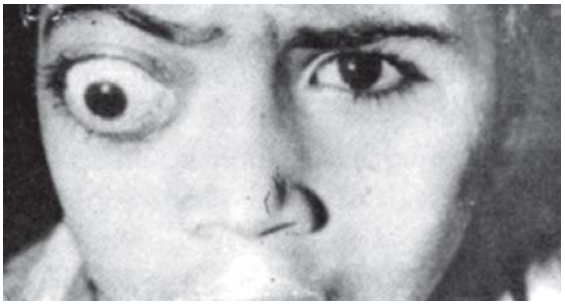


Fig. 26.10: Unilateral proptosis

Bilateral proptosis occurs generally due to endocrine disorders like thyrotoxic exophthalmos or thyrotropic exophthalmos (malignant ophthalmoplegia), developmental shallowing of the orbits, lymphomas and secondary deposits.

Proptosis in children occurs due to the causes that are quite different from those in adults. The common causes of proptosis in children include trauma, dermoid, hemangioma, optic nerve glioma, neurofibromatosis, rhabdomyosarcoma and leukemia.

Proptosis in adults may be due to orbital cavernous hemangioma, meningioma, mucocele of sinuses and secondaries from the breast, lungs and gastrointestinal tract.

Pulsating Exophthalmos

Etiology

When exophthalmos is due to an arteriovenous aneurysm, the angular vein and its tributaries near the inner canthus pulsate synchronously with the arterial pulse. It occurs after a severe blow or fall upon the head leading to a communication between the internal carotid artery and the cavernous sinus. The cerebral pulsation can also be transmitted to the bones of orbit or skull in patients with clefting defect causing meningocele, meningoencephalocele and neurofibromatosis.

Clinical Features

A constant rumbling of a waterfall is felt by the patient which can be heard on auscultation over the orbit by the examiner. The blood vessels of the conjunctiva and lids are dilated and the intraocular pressure is raised. The retinal signs include venous stasis, hemorrhages, exudates and optic disk edema.

Treatment

Ligation of the carotid artery may be tried after the location of shunt is determined on angiography.

Intermittent Proptosis

Etiology

Intermittent exophthalmos is caused by varicose dilatation of the orbital veins. It may occur following an orbital trauma or in patients of polycythemia vera.

Clinical Features

The exophthalmos is usually unilateral and becomes marked when the resistance to venous drainage is increased. Proptosis of varying degrees can occur on bending or screaming.

Diagnosis

The diagnosis of intermittent proptosis can be confirmed by ultrasonography using Valsalva maneuver. Carotid-cavernous fistula should be excluded.

Treatment

The surgical removal of varices is not without risk. Damage to neurovascular structures of the orbital cavity may occur during surgery.

Workup

History

A detailed history is helpful in establishing the cause of proptosis. The important points in history include:

1. Onset, course and duration of symptoms like pain, impairment of vision and diplopia.
2. Ocular or head injury.
3. Systemic diseases such as thyroid disorder, leukemia, sinus diseases and malignancy.
4. Congenital malformations.

Pseudoproptosis

Before proceeding for further evaluation, the causes of pseudoproptosis such as buphthalmos, axial high myopia, and retraction of upper eyelid must be excluded.

Clinical Examination

1. *Type of proptosis*: The type of proptosis, axial or nonaxial, should be ascertained. *Axial proptosis* is caused by a retrobulbar mass lesion in the muscle cone. *Nonaxial proptosis* is caused by a mass lesion outside the muscle cone. Direction of displacement of the globe is important. A superior displacement of the eye occurs in maxillary sinus growth while an inferomedial displacement results from lacrimal gland tumor, and inferolateral displacement from frontal or ethmoidal sinus lesions.
2. *Palpation*: Palpation around the eyeball may reveal the presence of a mass. Masses in the superonasal and superotemporal quadrants are not uncommon. Palpable mucocele, encephalocele and neurofibroma may be found in the superonasal quadrant. A dermoid or prolapsed lacrimal gland or lacrimal gland tumor can be palpable in the superotemporal quadrant.
3. *Pulsation*: Pulsations of the eye are caused by transmission of the vascular pulse. Neurofibroma and meningoencephalocele often produce pulsation without a bruit on auscultation but pulsation with or without bruit may be produced by carotid-cavernous fistula and arteriovenous communication. Dilated corkscrew epibulbar vessels may be found in arteriovenous malformation.
4. *Ocular movements*: Ocular movements can get restricted in a specific direction of gaze by an orbital mass. Bilateral thyroid ophthalmopathy involves multiple extraocular muscles and restricts the ocular movements. Graves ophthalmopathy most commonly involves the inferior rectus muscle causing its fibrosis and restriction of elevation.
5. *Eyelid abnormalities*: Some of the diseases causing proptosis may involve the eyelids. A strawberry birthmark in the skin of the eyelid may be seen in capillary hemangioma of the orbit. Plexiform neurofibroma causes an S-shaped curvature of the upper eyelid. Metastatic neuroblastoma may produce bilateral ecchymosis of the eyelids. Some abnormal movements of the eyelids have been recorded in Graves ophthalmopathy.
6. *Visual status*: Visual acuity, refraction and color vision should be recorded because a retrobulbar lesion can compress the optic nerve.
7. *Pupillary reaction*: The presence of a slow or nonreacting pupil is not rare in a case of proptosis. It indicates the optic nerve involvement.
8. *Intraocular pressure*: A rise of intraocular pressure may occur in Graves ophthalmopathy on attempted elevation. The intraocular pressure may also be raised in cavernous hemangioma and neurofibromatosis.
9. *Fundus examination*: Invasive meningioma may lead to papilledema or optic atrophy. Retinal striae are seen in cavernous hemangioma.

10. *Exophthalmometry*: Hertels exophthalmometer is generally used to measure the protrusion of the apex of cornea from the lateral orbital rim in both the eyes simultaneously. Exophthalmometric readings between 10 and 21 mm are considered normal. A difference of 2 mm between two eyes is considered abnormal.
11. *Examination of paranasal sinuses:* Paranasal sinuses should be examined to exclude a mass or a mucocele encroaching on the orbital cavity.
12. *Systemic examination*: It should be conducted to exclude the developmental anomalies of the orbit, neuroblastoma and leukemia especially in children, and thyroid disorder (Graves disease) and malignancies of breast, lungs and gastrointestinal tract in adults.
13. *Cranial nerves*: Examination of cranial nerves especially II, III, IV, V, VI, VII and VIII are important since they may get involved in the inflammatory diseases of the orbit and cavernous sinus, and Wegener's granulomatosis and neoplasm.

Investigations

1. *Computed tomography (CT)*: CT scan is the most valuable technique for delineating the shape, extent, location and character of an orbital lesion. CT scan is indicated in most cases of proptosis because it provides a good view of bony orbit, extraocular muscles and retained metallic foreign body. Spiral CT scan is indicated for children.
2. *Magnetic resonance imaging (MRI)*: MRI is a noninvasive imaging technique which provides the best anatomical details of the orbit. It gives better soft tissue details and view of the orbital apex density. In spite of these advantages, MRI is contraindicated in patients with a retained intraocular or orbital metallic foreign body.
3. *Ultrasonography*: Standardized A-scan provides unidimensional images of the orbital soft tissues by a series of spikes of varying heights depending on the echogenic characteristics of each tissue. B-scan ultrasonography presents two-dimensional images and helps in identifying the size, shape and position of an orbital lesion.
4. *Venography and arteriography*: Before the introduction of CT scan and MRI, orbital venography was used in the diagnosis of orbital varices but now it is rarely performed. Arteriography is indicated in the diagnosis of aneurysm and arteriovenous malformation. The technique carries a risk of neurological and vascular complications. Magnetic resonance angiography is a recent noninvasive technique which allows the visualization of large and medium sized vessels.
5. *Laboratory studies*: All patients with thyroid ophthalmopathy should be screened for serum T3, T4 and thyroid-stimulating hormone (TSH) estimation. These tests are abnormal in nearly 90% of patients. Estimation of serum angiotensin-converting enzyme may be helpful in the diagnosis of sarcoidosis.
6. *Biopsy*: The accurate diagnosis of an orbital mass lesion requires a histopathological examination. Fine needle aspiration biopsy is a simple technique but may not allow sufficient biopsy specimen to permit a firm diagnosis. Excisional biopsy is obtained through orbitotomy. Frozen-section analysis ensures a complete tumor removal. Cell-marker studies are required for the orbital lymphoid lesions.

ENOPHTHALMOS

Recession (inward displacement) of the eyeball within the orbital cavity is called *enophthalmos.* Enophthalmos may be unilateral or bilateral. It must be differentiated from microphthalmos and phthisis bulbi. Common causes of enophthalmos are listed below.

1. Blow-out fracture of the orbit unassociated with orbital hematoma (most common).

2. Atrophy of the orbital tissue due to senility, dehydration, after repeated periocular injections of corticosteroids and following irradiation of the orbit.
3. Neurogenic causes include Horner's syndrome and paralysis of superior and inferior oblique muscles.

TUMORS OF THE ORBIT

Orbital tumors are not common. They can be classified as benign and malignant or as primary, secondary and metastatic. A simple classification of common orbital tumors is given in Table 26.4.

Benign Orbital Tumors

Dermoid

Dermoid is the most common congenital orbital tumor frequently found adjacent to the frontozygomatic suture (Fig. 26.11). Globe may be displaced due to progressive proptosis. Hair, cartilage, teeth, bone and other tissues may be found in the dermoid.

Table 26.4: Different types of orbital tumors

Primary Orbital Tumors	
Congenital	Dermoid, Dermolipoma, Teratoma
Vascular	Hemangioma
Neural	Optic nerve glioma, Neurofibroma, Nerofibromatosis, Meningioma
Mesenchymal	Rhabdomyosarcoma
Lymphoid	Lymphoma
Lacrimal	Pleomorphic adenoma, Malignant mixed tumor
Secondary Orbital Tumors	
Eye	Retinoblastoma, Malignant melanoma
Eyelids	Adenocarcinoma, Squamous cell carcinoma
Sinuses	Squamous cell carcinoma of maxillary sinus and nasopharynx
Brain	Meningioma
Metastatic Orbital Tumors	
Children	Neuroblastoma, Leukemia
Adult	Carcinoma of breast and lungs, Cutaneous malignant melanoma

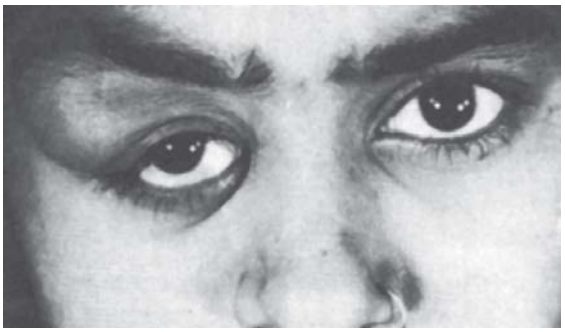


Fig. 26.11: Orbital dermoid

Lipodermoid

Lipodermoid is a solid tumor which often occurs beneath the conjunctiva near the superotemporal quadrant of orbit. Most lipodermoids do not need any treatment.

Hemangioma

Hemangiomas are benign tumors of the orbit which manifest in two forms—capillary and cavernous.

Capillary hemangioma is a common orbital tumor of childhood. The tumor involves the skin producing an elevated strawberry discoloration. Deep seated orbital hemangioma causes bluish discoloration. The capillary hemangioma usually involves the superonasal quadrant of the orbit and the medial part of upper eyelid. The tumor causes cosmetic deformities of the eyelid and leads to anisometropia, strabismus and deprivation amblyopia.

Treatment includes local corticosteroids injection of an equal mixture of betamethasone, 6 mg/ml and triamcinolone, 40 mg/ml. Repeat injection may be needed in some patients. Small lesions can be excised surgically. Radiation therapy has potential side effects.

Cavernous hemangioma (Fig. 26.12) is the most common benign tumor of orbit in adults. It may cause slowly progressive proptosis, hyper-

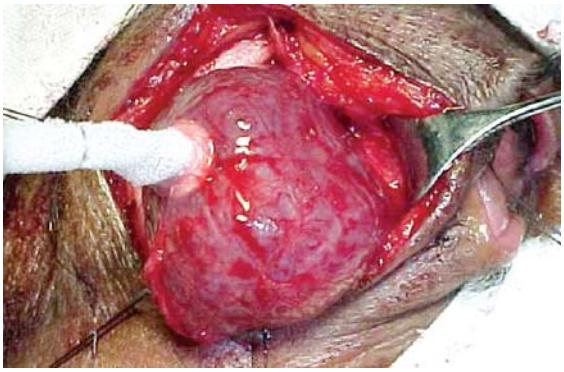


Fig. 26.12: Cavernous hemangioma during orbitotomy (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

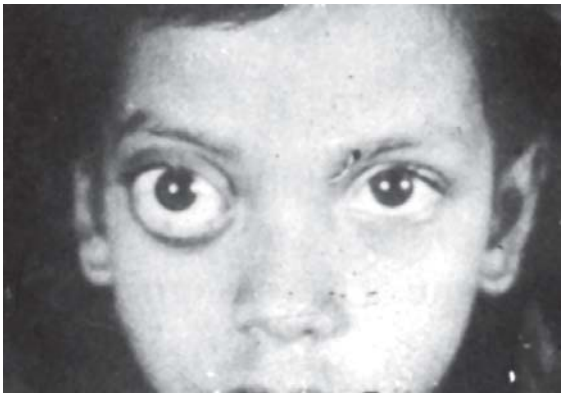


Fig. 26.13: Glioma of optic nerve

metropia, optic nerve compression, retinal striae, raised intraocular pressure and strabismus. CT scan may localize the tumor in the orbit. The surgical excision of the tumor can be done through lateral orbitotomy.

Glioma

Glioma may be associated with neurofibromatosis (25 to 60%). It occurs during the first decade of life and causes early visual impairment. Glioma produces a painless, unilateral axial proptosis (Fig. 26.13). Strabismus, papilledema and optic atrophy are other clinical features. Chiasma may be involved in nearly 50% of cases of optic nerve glioma. CT scan and MRI show a characteristic fusiform enlargement of the optic nerve (Fig. 26.14).

Patients with good vision should be followed up without any treatment. Surgical excision, radiotherapy and chemotherapy are the treatment modalities for the tumor causing visual loss.

Neurofibroma

Neurofibromas are mainly composed of proliferating Schwann cells within the nerve sheath. Neurofibromatosis (von Recklinghausen's disease) is a type of phacomatosis which often involves the orbit. The disease is characterized by

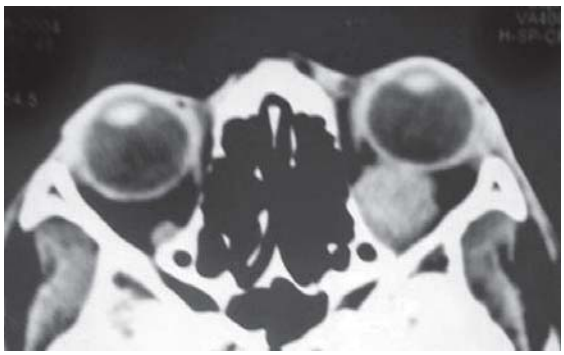


Fig. 26.14: CT scan of orbit showing spindle-shaped dilatation of optic nerve (*Courtesy*: Dr MS Bajaj, Dr RP Centre, New Delhi)

the presence of plexiform neurofibromas at the lateral part of the upper eyelid giving an S-shaped contour to the eyelid, pulsating exophthalmos and congenital glaucoma. Neurofibromatosis may be associated with optic nerve glioma.

Meningioma

Meningioma arises from the arachnoid villi and is usually attached to the dura mater. The sphenoidal ridge meningioma usually originates from the lesser wing of the sphenoid bone and extends into the orbit. CT scan shows a diffuse thickening of the optic nerve (Fig. 26.15) and a localized bone thickening associated with

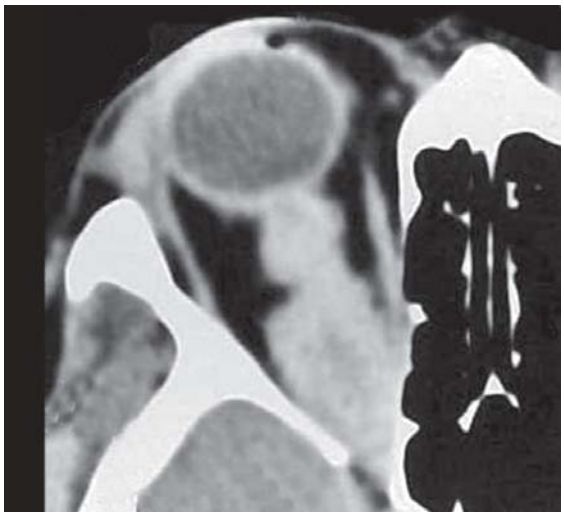


Fig. 26.15: CT scan shows a diffuse thickening of optic nerve characteristic of optic nerve sheath meningioma (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

abnormal calcification. Radiotherapy stabilizes the condition. Surgery is indicated if there is tendency for an intracranial extension.

Malignant Orbital Tumors

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common primary malignant orbital tumor of childhood (approximately 87% before 15 years of age). It is a highly malignant neoplasm arising from undifferentiated pluripotent mesenchymal elements in the orbital soft tissue and not from extraocular muscles.

Rhabdomyosarcoma produces a rapidly progressive unilateral proptosis (Fig. 26.16) of sudden onset which may be associated with ptosis and strabismus. The tumor typically involves the superonasal quadrant, but it may invade any part of the orbit.

CT scan, MRI and ultrasonography may help to define the site and extension of the neoplasm. An excisional biopsy should be performed through anterior orbitotomy. The treatment of rhabdomyosarcoma is local irradiation combined with chemotherapy. A total of 4500-6000 cGys of irradiation is given in 6 weeks.

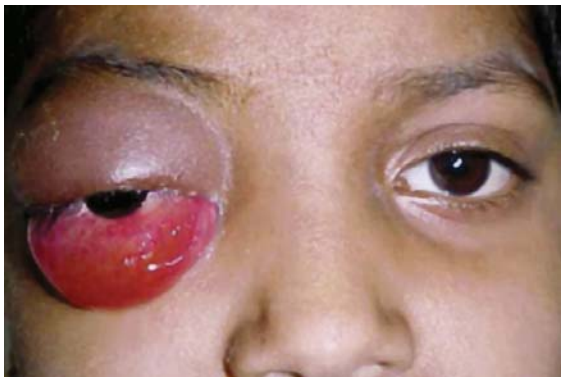


Fig. 26.16: Rhabdomyosarcoma of orbit (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

Lymphoma

Lymphoma of the orbit can occur either in isolation or as a part of systemic disease. They account for nearly 20% of all orbital neoplasms. Lymphoid tumors can be divided into two groups—benign reactive lymphoid hyperplasia and malignant lymphomas (90%).

Orbital lymphoma (Fig. 26.17) presents as a painless progressive palpable mass in the anterior orbit below the conjunctiva. It is more common in elderly females (50-70 years of age). Fifty percent of patients with orbital lymphoma eventually develop the systemic disease.

CT scan, tissue biopsy and immunohistochemical studies help in establishing the diagnosis. The reactive infiltrates demonstrate follicular hyperplasia without a clonal population of cells. Orbital lymphoma shows monoclonality or cytologic and architectural atypia. Malignant lymphoma have multiclonal infiltrates.

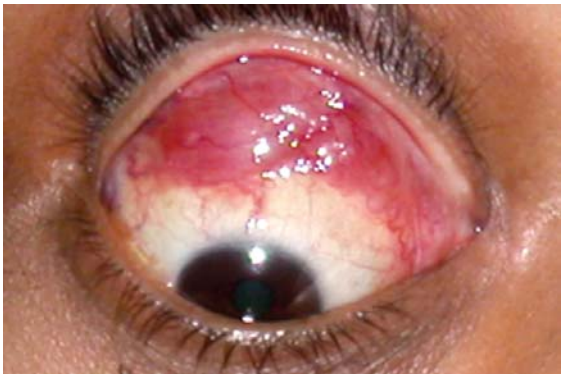


Fig. 26.17: Orbital lymphoma presenting with salmon pink mass in upper conjunctiva (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

Fig. 26.18: Bilateral ecchymotic proptosis in a child from metastatic neuroblastoma (*Courtesy*: Dr SG Honavar, LVPEI, Hyderabad)

Radiotherapy is the treatment of choice. Eye should be protected by metallic contact lens and a dose of 2000-2500 cGys is recommended. More aggressive lymphomas need chemotherapy or a combination of radiotherapy and chemotherapy.

Secondary Orbital Tumors

Tumors from contiguous structures such as the eyeball, eyelids, sinuses and brain may invade the orbit. Retinoblastoma and malignant melanoma of choroid often invade the orbit during the stage of extraocular extension. Squamous cell carcinoma and adenocarcinoma of the eyelid can secondarily invade the orbit.

Both epithelial and nonepithelial tumors from the nose and paranasal sinuses commonly invade the orbit. The primary tumor arises usually within the maxillary sinus or the nasopharynx causing epiphora, epistaxis or nasal obstruction.

Metastasis in the Orbit

Neuroblastoma and leukemia often metastasize to the orbit of children.

In *neuroblastoma* the primary tumor may be in the abdomen, the mediastinum or the neck. The metastatic neuroblastoma causes bilateral ecchymotic proptosis (Fig. 26.18) and Horner's syndrome.

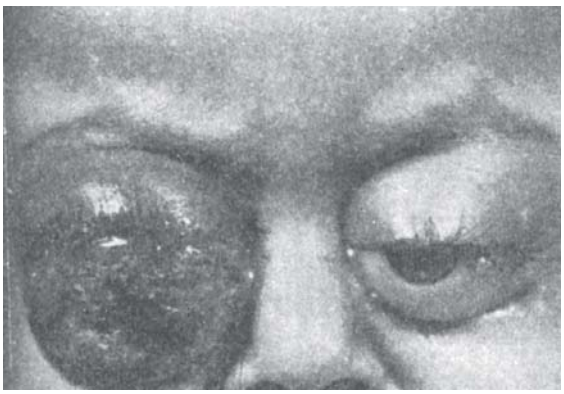


Fig. 26.19: Bilateral leukemic proptosis

Acute lymphoblastic leukemia can metastasize to the orbit producing a unilateral or a bilateral proptosis (Fig. 26.19). Rarely, a leukemic orbital mass, *chloroma*, is found in acute myeloid leukemia which represents a solid collection of immature malignant white blood cells.

Carcinoma of the breast and lungs, and cutaneous malignant melanoma can metastasize to the orbit of adults. The common source of orbital

metastasis in women is carcinoma of the breast, while in men it is bronchogenic carcinoma.

Pain, proptosis, local bone destruction and ophthalmoplegia are presenting features of the metastasis in the orbit. CT scan, elevation of serum carcinoembryonic antigen level and fine needle aspiration biopsy are helpful in the diagnosis. The treatment includes local radiation therapy.

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CHAPTER

27 Operations upon the Eyeball and its Adnexa

ANESTHESIA

Operations upon the eyeball can be performed either under local or general anesthesia. Most ophthalmic surgeons prefer local anesthesia as it is safe and free from the after-effects of general anesthesia. Success of an intraocular surgery largely depends upon a perfect anesthesia and akinesia. Preoperative administration of systemic sedatives such as alprazolam (0.50-1 mg) and a nonsteroidal anti-inflammatory drug 2 hours before operation allays the apprehension of the patient, while retaining his cooperation during surgery.

The general anesthesia is required for complicated vitreoretinal surgeries, uncooperative patients and children. Rise of intraocular pressure and increased oozing of blood intraoperatively, and postoperative cough, vomiting and straining are the common hazards of general anesthesia.

Local Anesthesia

The local anesthesia consists of both surface and infiltration anesthesia. The conjunctiva and the cornea are anesthetized by topical application of 2% or 4% xylocaine or 0.50% proparacaine two to three times at an interval of 5 to 10 minutes.

The infiltration anesthesia is obtained by local infiltration of 2% xylocaine. Ciliary ganglion and its surrounding structures are anesthetized by retrobulbar block. It is achieved by injecting 1 ml of 2% xylocaine with a fine long needle (5 cm) at the outer and lower angle of the orbit directed towards the apex of the orbit. This block causes a temporary paralysis of extraocular muscles and slight dilatation of pupil.

The retrobulbar injection of local anesthetic agents is not a benign procedure. Mild to severe retrobulbar hemorrhage warranting postponement of operation, perforation of globe, injury to the optic nerve, meningeal irritation and respiratory distress are the potential complications. These complications may be reduced by giving a peribulbar anesthesia.

The peribulbar anesthesia is safer than retrobulbar anesthesia because the anesthetic agent is injected outside the muscle cone and thus chances of intraocular and optic nerve injuries are minimized (Fig. 27.1). The injection is given through the lower eyelid at the junction of medial two-thirds and lateral one-third of the inferior orbital margin. 4 to 6 ml of the solution is injected just posterior to the equator. A firm pressure by fingers or a superpinkie ball is usually applied for 20 minutes. The efficacy of the block is then evaluated. Incomplete block requires an additional injection of 2 ml of anesthetic agent either given at the same site or through the upper lid superonasally at the junction of medial one-third and lateral two-thirds. The anesthesia does not begin as rapidly as the

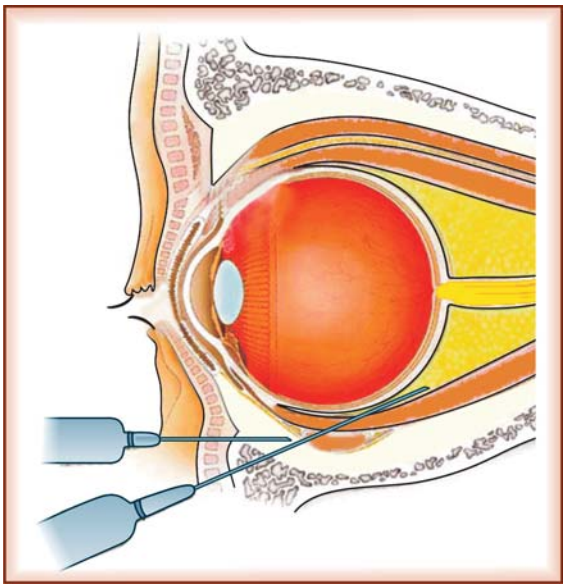


Fig. 27.1: In peribulbar anesthesia the needle remains in peripheral space and in retrobulbar block it lies in muscle cone

retrobulbar block and about 10% cases may need an additional anesthesia. However, it is a simple and safe technique which also eliminates the facial block.

Akinesia

Akinesia during an ocular surgery can be obtained by the facial block which causes a temporary paralysis of orbicularis oculi. One of the two common techniques, O'Brien's and Van Lint's, may be employed.

In *O'Brien's technique* the patient is asked to open and close his mouth and the position of the mandibular condyle is ascertained. About 5 ml of xylocaine is injected down to the periosteum covering the neck of the mandible and local massage is applied.

In *Van Lint's technique* 3 to 4 ml of xylocaine is infiltrated through a needle inserted up to the periosteum of the malar bone, at a point about 1 cm below and behind the outer canthus, and then pushed upward towards the temporal fossa, downward towards the infraorbital foramen and towards the tragus. The branches of the facial nerve are blocked as they course over the malar bone.

Sometimes, the superior rectus muscle may continue to pull the eyeball upwards despite the peribulbar injection. Infiltration of 0.5 ml of xylocaine 3 mm behind its insertion produces a transient paralysis of the muscle.

With the introduction of operating microscope (Fig. 27.2), there is an increasing trend to perform most of the ocular surgeries under the microscope.

SURGERY ON THE LID

Surgery on eyelid is done for hordeolum externum, chalazion, spastic and cicatricial entropion, ectropion and ptosis.

Stye

Hordeolum externum generally subsides with frequent hot compresses and application of an

Fig. 27.2: Operating microscope (*Courtesy*: Carl Zeiss India)

antibiotic ointment. When pus is pointing and the condition not resolving, a small incision is made over the pus point after giving local anesthesia. The wound is dressed with an antibiotic ointment and a bandage can be applied. The bandage may be discarded after a few hours.

Chalazion

Chalazion can be operated either from the conjunctival or from the skin side. Infiltration anesthesia is given in the area of chalazion. A chalazion clamp is applied and the lid is everted. A vertical incision is made in the conjunctiva (Fig. 27.3) and the cheesy material oozes out. The cavity is curetted by a chalazion scoop. If there is a thick granulation tissue, it should be removed with scissors. Small incision does not require suturing. Antibiotic ointment is applied and the eye is bandaged for a few hours. If chalazion has become subcutaneous, it can be incised from the skin side by giving a horizontal incision.

Entropion

Spastic entropion of the lower lid can be corrected by removal of a small strip of the skin and orbicularis oculi muscle. Permanent relief may be obtained by modified Wheeler's operation in which a band of orbicularis is exposed, divided in the center and the flaps are overlapped for 4 to 5 mm.

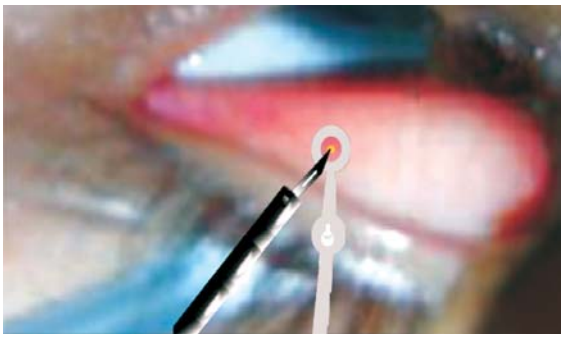


Fig. 27.3: Incision of chalazion from conjunctival side

Cicatricial entropion is usually relieved by plastic operations. The various operations are based on the following principles:

1. Correction of the misdirection of eyelashes
2. Transplanting the lashes to a higher level, and
3. Correction of the distorted tarsal plate.

In *Burrow's operation (Tarsal fracture)* a horizontal incision in the sulcus subtarsalis passing completely through the whole thickness of the lid (video) excepting the skin relieves the internal pull from the cicatricial tissue (Fig. 27.4A). The temporal end of intermarginal strip is divided by a full-thickness vertical incision.

Jaesche-Arlt operation combines the principles of correction of the misdirection of lashes by splitting the lid along the gray line and transplantation of the zone of hair follicles to a slightly higher position by removal of a crescentic piece of the lid skin.

Snellen's partial tarsectomy is advocated in entropion associated with a distorted tarsal plate (video). Nearly two-thirds thickness of the entire upper tarsal plate is dissected and removed and skin-muscle-tarsal plate and tarsal plate-muscle-skin sutures are applied to close the wound (Fig. 27.4B).

Jones procedure (Fig. 27.4C) corrects involutional entropion (video).

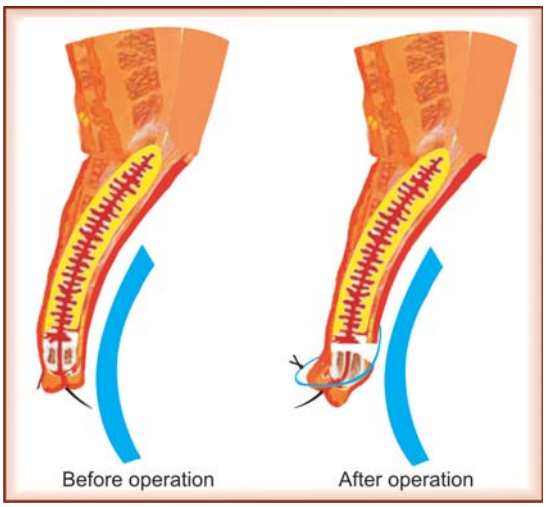


Fig. 27.4A: Burrow's operation (Tarsal fracture)

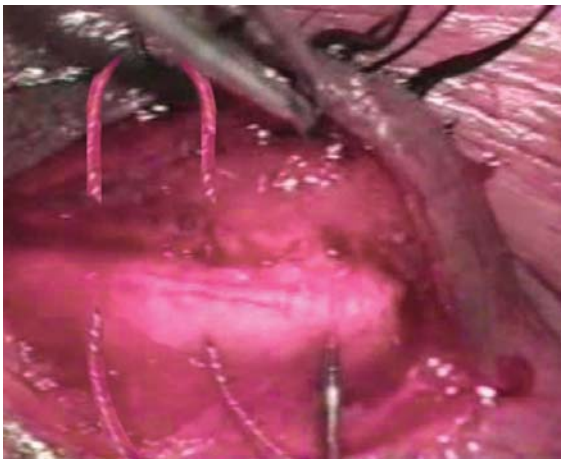


Fig. 27.4B: Snellen's partial tarsectomy

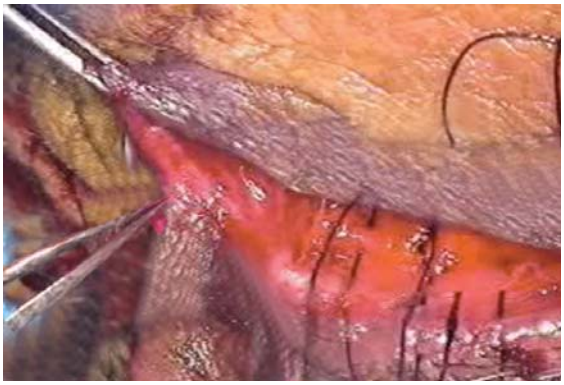


Fig. 27.4C: Jones procedure

Ectropion

Senile ectropion of the lower lid is usually corrected by *lateral tarsal strip procedure and conjunctivoplasty (video)*.

For correcting the cicatricial ectropion several operations are devised. The principles governing these operations are:

1. Shortening the palpebral aperture
2. Shortening the lid and transplanting it to a higher level, and
3. Removal of the contracture.

Byron-Smith modification of Kuhnt-Szymanowski operation is a procedure wherein a triangular piece of the conjunctiva and the tarsus is excised from the middle part of the lower lid and the lid is split along the gray line from the triangle to the outer canthus. An appropriate triangular piece of the skin is removed at the outer canthus. The gap in the lower lid is closed. Then the lid margin is mobilized upwards and outwards to cover the skin incision (video).

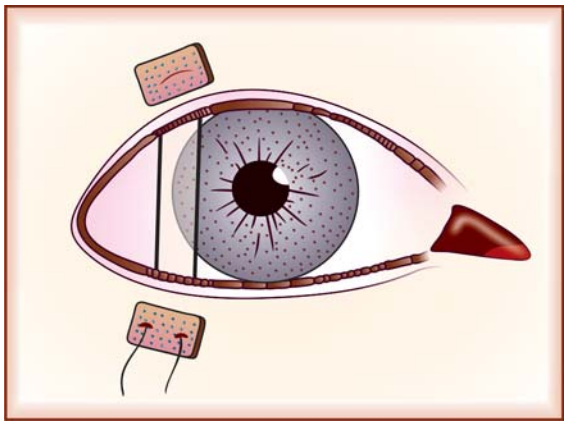


Fig. 27.5: Lateral tarsorrhaphy

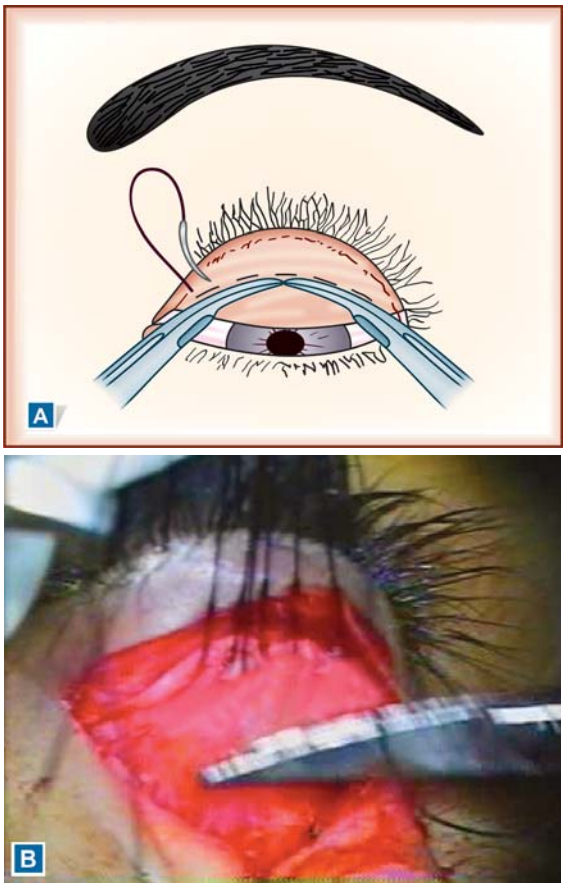
Wharton-Jone's V-Y operation is indicated for mild type of ectropion of the lower lid. The scar in the lower lid is removed by a V-shaped incision having its apex away from the lid margin. The skin is undermined and sutured in a Y-shaped manner.

Lateral tarsorrhaphy (Fig. 27.5), a procedure in which the palpebral aperture is shortened by sewing the lids at outer canthus, is beneficial in paralytic ectropion. Blepharoplasty is indicated in extensive cicatricial ectropion.

Ptosis

Before any ptosis surgery, the action of levator palpebrae superioris (LPS) and other extrinsic muscles particularly the superior rectus, is carefully assessed, and depending on it one of the following operative procedures may be utilized.

Fasanella-Servat operation is indicated in mild degree of ptosis with good levator function. The lid is everted and two hemostats are applied. The upper tarsal plate alongwith the LPS, Muller's



Figs 27.6A and B: (A) Fasanella-Servat operation, (B) Everbusch's operation

muscle and the conjunctiva are excised (Fig. 27.6A) and a running suture is placed (video).

If the LPS action is weak and other extraocular muscles are normal, the ptosis is corrected by shortening the levator. The muscle can be resected either through the conjunctiva (Blaskowics' operation) or through the skin route (Everbusch's operation).

In *Blaskowics' operation* the lid is everted on a spatula and an incision is made at the upper border of the tarsal plate. The conjunctiva is reflected and the levator tendon is exposed. The tendon is freed from its attachment and the upper strip of tarsal plate is excised. Three double-armed sutures are passed through the tendon 18 to 20 mm above its insertion. The tendon is then cut distal to the sutures and the cut end is anchored to the tarsal plate 2 to 3 mm above the lash margin. Another set of double-armed sutures is threaded through the tendon 3 mm above the line of its present attachment, the sutures are brought out through the skin and tied just midway between the upper and lower limits of the lids to make the natural lid folds.

In *Everbusch's operation* a better exposure of LPS through skin approach can be obtained (Fig. 27.6B) and a large resection of muscle (22-24 mm) may be performed.

Motais' operation is indicated when the levator is completely paralyzed and the superior rectus muscle is functioning normally. The middle-third of the tendon of the superior rectus is transplanted to the upper border of the tarsal plate through a subconjunctival approach. The operation must not be performed in unilateral cases as it causes a varying degree of vertical muscle imbalance.

Hess' operation or *frontalis sling operation* is a better choice than Motais' operation in the patients with poor LPS function wherein the elevating action of frontalis muscle is utilized for the correction of ptosis (video).

The lid is anchored to the frontalis muscle using a sling of supramyd. Three small incisions in the upper lid 3 mm above the lid margin, two incisions 5 mm above the medial and the lateral part of the eyebrow, and one incision 15 to 16 mm above and between the two are made. Supramyd or 3-0 polypropylene suture or fascial strips are passed through the openings in the lid, then through the openings above the eyebrow (video). The one end of the sling is cut and secured by sutures and the other passes through the top incision from either side of the brow. The sling must be drawn tightly to obtain a full correction (Fig. 27.7).

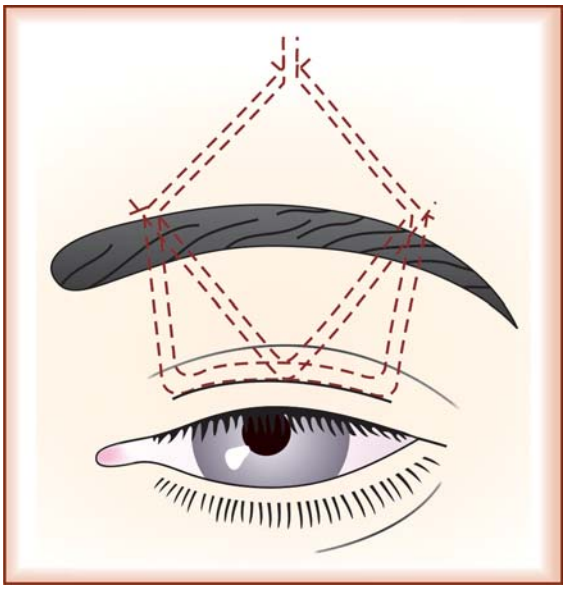


Fig. 27.7: Frontalis sling operation

SURGERY ON THE CONJUNCTIVA

Surgery on the conjunctival sac is performed under topical or infiltrative anesthesia where 2% xylocaine with adrenaline is injected in the subconjunctival space.

Peritomy

Persistent progressive vascularization of the cornea may be controlled by removal of a strip of conjunctiva 2 mm in width from the limbal area. The vessels should be cauterized.

Pterygium Surgery

A stationary thin pale pterygium seldom warrants excision. Pterygium needs removal when it is progressing and causing cosmetic disfigurement. If pterygium has already invaded the pupillary area it is advisable to wait till it crosses the area as the removal of the apex of the pterygium leaves a thick scar.

Operation for pterygium is performed under local anesthesia. The neck of the pterygium is lifted with a toothed forceps and it is shaved from the cornea with a knife. The body of pterygium is freed from the sclera and excised by giving two converging incisions by the scissors. The exposed sclera may be either covered by mobilizing the conjunctiva or left bare especially near the limbus.

In *D'Ombren's operation* or *bare sclera technique* the pterygium is lifted by a forceps and a 5 mm incision is made diverging from the limbus. The subepithelial degenerative tissue is thoroughly dissected and the head, neck and about 2 mm of the body of the pterygium is excised in one triangular piece leaving an exposed area of the sclera, approximately 4 mm wide. The conjunctiva is sutured.

In *McReynold's operation* the apex of the pterygium is lifted and a double-armed suture passed in it. Then a pocket is made in the upper fornix and the head of the pterygium is buried and sutured in the pocket. This surgery is not done now-a-days.

The current techniques for the management of pterygium include a *conjunctival autograft* from the same or the opposite eye (video) or an *amniotic membrane transplantation* (video) after the excision of pterygium.

OPERATIONS UPON THE CORNEA

Keratectomy

Superficial irregular corneal scars are shaved off with a diamond knife or using an excimer laser in keratectomy and an amniotic membrane transplant is given (video). The corneal epithelium grows over the defect. Keratectomy is indicated in recurrent corneal erosions, filamentary keratitis, and multiple embedded foreign bodies in the cornea.

Keratoplasty

In keratoplasty or corneal transplantation, the opaque corneal disk (Fig. 27.8A) is replaced by a corresponding sized graft taken from the healthy cornea of a donor. The keratoplasty is usually of two types—lamellar (partial-thickness graft) and penetrating (full-thickness graft).

Indications

The indications for lamellar keratoplasty include superficial corneal scars, stromal corneal dystrophies and recurrent pterygia.

The indications for penetrating keratoplasty (PKP) are as follows:

1. *Optical*: Keratoconus, deep corneal dystrophies, bullous keratopathy.
2. *Therapeutic*: To prevent the impending perforation of cornea.
3. *Tectonic*: To restore the cornea after perforation.
4. *Cosmetic*: To improve the look of the eye even though it is blind.

Procedure

The donor cornea can be obtained from an eye bank wherein short-term (McCarey-Kaufman medium or MK medium) or long-term (organ culture) corneal preservation facilities are available. Freshly procured cornea (2-4 hours) often gives good results.

The operation can be performed either under local or general anesthesia. The size of the graft is determined, the grafts smaller than 6 mm are inadequate while grafts larger than 8.5 mm are prone for complications. The operation is performed under following steps (video).

1. The corneal button is cut, using a trephine, from a donor corneoscleral ring.
2. The host corneal button is excised using the corneal trephine and scissors.
3. The donor corneal button is sutured in the host cornea.

Complications

The penetrating keratoplasty is not free from complications which include anterior synechia, secondary glaucoma and graft rejection. In spite of these complications, the results of keratoplasty are gratifying in restoring the vision (Fig. 27.8B). In failed keratoplasty (Fig. 27.8C) regrafting may be performed after nine months of the first graft.

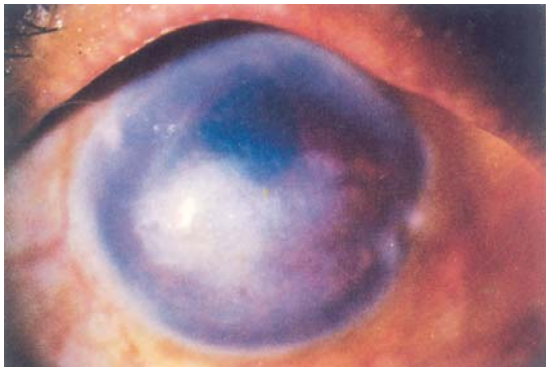


Fig. 27.8A: Opaque cornea

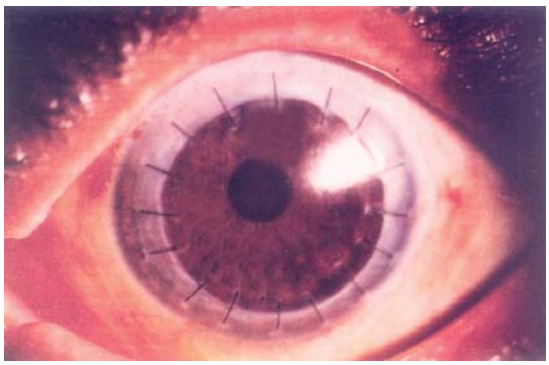


Fig. 27.8B: Successful corneal graft following PKP (*Courtesy*: Dr MK Aasuri,LV Prasad Eye Institute, Hyderabad)

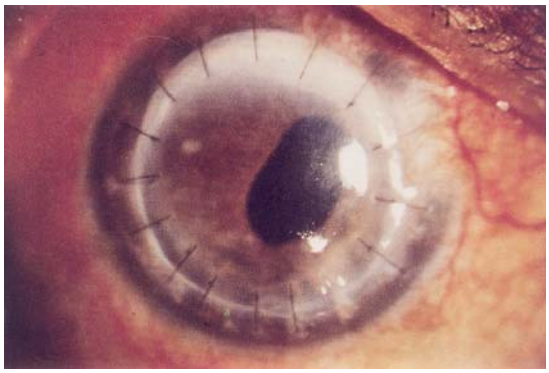


Fig. 27.8C: Failed corneal graft (*Courtesy*: Dr MK Aasuri, LVPEI, Hyderabad)

Refractive Surgery

Surgical interventions for the correction of the refractive errors have become popular of late. Different surgical techniques are being used to correct myopia and hypermetropia.

Surgical Correction of Myopia

1. *Radial keratotomy* (RK) corrects the myopia in the range of 2 to 6 D. In this technique a central corneal optical zone (3-4 mm) is spared and 8 or 16 radial corneal incisions are placed (Fig. 27.9). Lack of predictability of results and glare are its drawbacks.
2. *Photorefractive keratectomy* (PRK) ablates the central anterior corneal stroma with the help of an excimer laser (193 nm) to reduce the corneal curvature. Corneal haze and glare may occur as complications.
3. *Phakic lens implantation* involves the placement of an anterior chamber lens in a phakic eye. It is a satisfactory procedure to correct high degree of myopia.
4. *Laser-in-situ keratomileusis* (LASIK) is an effective surgical technique for the correction of myopia. A hinged corneal flap is prepared by using a microkeratome. The refractive keratectomy (in corneal stroma) is performed using an excimer laser. Then the flap is replaced (video).

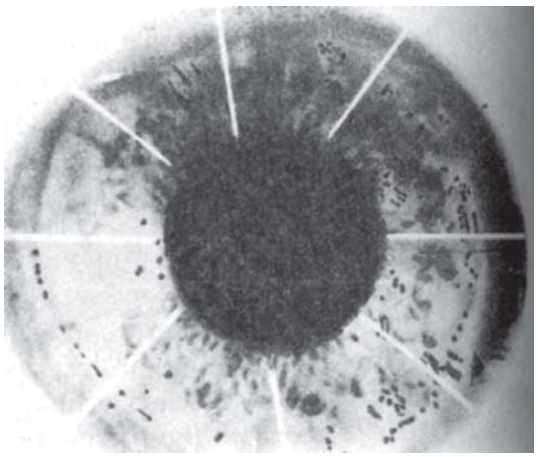


Fig. 27.9: Radial keratotomy

Surgical Correction of Hypermetropia

1. *Laser thermal keratoplasty (LTK)* involves the use of holmium laser to reshape the cornea for correction of hypermetropia. The holmium laser is an infrared laser that shrinks the corneal stromal collagen fibers.
2. *Intraocular lens implantation*, at the time of cataract surgery, is the most popular and safe method to correct aphakic hypermetropia. Suitable cases of aphakia can be managed by secondary lens implantation.

PARACENTESIS

Paracentesis is indicated in secondary glaucoma, massive hypopyon and hyphema. After local anesthesia, the eyeball is steadied with a fixation forceps and a small incision is made nearly 2 mm within the limbus with a keratome or a paracentesis needle. The posterior lip of the wound is depressed by an iris repositor so that the aqueous or blood or pus escapes slowly from the anterior chamber.

OPERATIONS UPON THE IRIS

Iridectomy

Iridectomy is an abscission of a part of the iris. It is performed for the following purposes.

1. Removal of prolapsed iris
2. For optical purpose (*optical iridectomy*)
3. As a part of cataract operation
4. As a part of glaucoma operation
5. For removal of foreign body, cyst or tumor of the iris.

A prolapsed iris during cataract extraction is usually reposited. However, postoperative or posttraumatic prolapsed iris should not be

reposited as it carries the risk of intraocular infection. A small prolapse is cauterized but an extensive one needs abscission. For iridectomy the previous section is opened with the help of an iris repositor. The iris is grasped with a forceps and drawn through the incision and abscised with de Wecker's or Vanna's scissors. The pillars are reposited with the iris repositor and the wound is closed by end-to-end sutures.

Optical iridectomy was used to be performed to obtain visual improvement in central corneal leukoma with a clear periphery, and zonular or central nonprogressive cataract. Subjects for iridectomy should be investigated with the help of a stenopeic-slit placed in front of a dilated pupil to find the site for best visual acuity. Generally, the site of selection for optical iridectomy is inferonasal quadrant for those who are engaged in near work and inferotemporal for those involved in outdoor activity. Ideally, an optical iridectomy should be as narrow as possible to avoid glare.

Iridectomy for Occlusio Pupillae and Updrawn Pupil

A preliminary sector iridectomy is occasionally performed in complicated cataract and subsequently the cataract is extracted. Iridectomy is also performed to reform the pupil in occlusio pupillae or in postoperative extreme updrawn pupil. In some cases, along with the iris, the remnants of lens capsule and organized blood clot are also abscised, the procedure is known as *Elschnig's operation of capsulo-iridotomy*.

Iridotomy

Surgical iridotomy is employed for prevention of iris bombé. Since the iris is an elastic tissue, a small surgical cut by scissors causes its retraction and eventual formation of an opening. Laser iridotomy is done for the management of primary angle-closure glaucoma.

SURGERY FOR DEVELOPMENTAL GLAUCOMA

Developmental glaucoma is usually managed by surgery. Goniotomy and trabeculotomy are effective procedures in congenital and infantile glaucoma.The results of surgery are excellent when operation is performed prior to the development of buphthalmos. Long-standing cases of developmental glaucoma with a corneal diameter of 15 mm or more are unsuitable for operative interference. The traditional operation of choice for developmental glaucoma has been goniotomy. The operation is aimed to slit open the mesodermal remnants of anterior chamber so as to permit the usual drainage of the aqueous through the trabeculum.

Goniotomy

Goniotomy is done under general anesthesia. The anterior chamber is filled with viscoelastic. Contact lens is applied, the goniotomy knife is passed across the anterior chamber to the trabeculum and an arcuate incision of about 120° is made in the anterior aspect of the meshwork by the tip of the knife. More than one goniotomy is required in different segments of the angle of the anterior chamber to normalize the intraocular pressure (IOP). Hyphema and postoperative rise in IOP may occur as complications.

Trabeculotomy

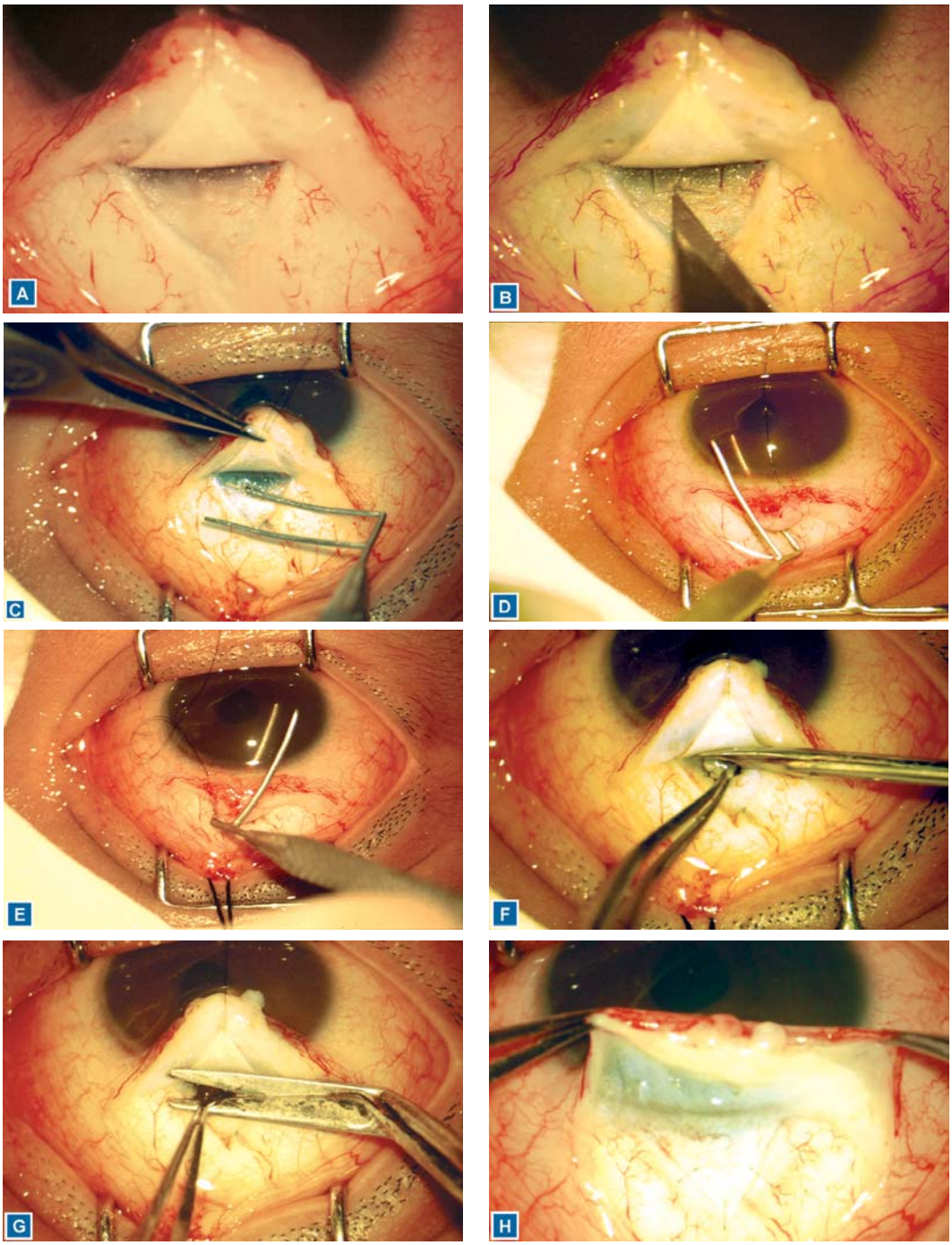
Trabeculotomy gives better results than goniotomy in developmental glaucoma.

Indications

Trabeculotomy is indicated in following conditions:

1. Failed cases of goniotomy, and
2. When the corneal clouding prevents goniotomy.

The operative steps of trabeculotomy are given below (Figs 27.10A to H):



Figs 27.10A to H: Steps of trabeculotomy: (A) Triangular-shaped partial thickness scleral flap, (B) Schlemm's canal explored, (C) Trabeculotome introduced into the Schlemm's canal, (D) Trabeculotome introduced into the anterior chamber, (E) Trabeculotome rotated on the other side, (F) Trabecular block excised, (G) Iridectomy done, (H) Closure of the scleral incision (*Courtesy*: Dr AK Mandal, LVPEI, Hyderabad)

1. A limbal-based conjunctival flap is prepared.
2. A triangular-shaped partial thickness scleral flap is dissected (Fig. 27.10A).
3. A radial incision is made across the scleral spur to cut the outer wall of Schlemm's canal (Fig. 27.10B).
4. A trabeculotome is introduced into the canal (Fig. 27.10C) and rotated into the anterior chamber (Fig. 27.10D) to disrupt the internal wall of the canal and the trabecular meshwork. The same procedure is repeated on the other side of the incision (Fig. 27.10E) rupturing a total of about 120° of the trabecular meshwork.
5. A small block of deep trabecular tissue is excised (Fig. 27.10F).
6. A peripheral iridectomy is performed (Fig. 27.10G).
7. The scleral flap is reposited and sutured (Fig. 27.10H).
8. The conjunctival flap is closed with running sutures.

If trabeculotomy fails then a trabeculectomy may be performed.

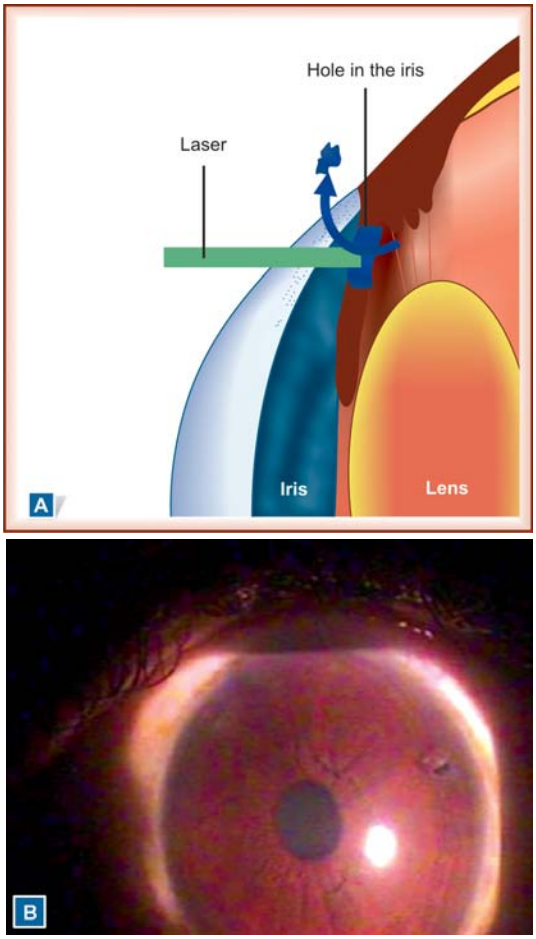
SURGERY FOR PRIMARY ANGLE-CLOSURE GLAUCOMA

The primary angle-closure glaucoma (PACG) is managed surgically and medical treatment is used only to reduce the pressure during the acute angle-closure attack.

The glaucoma operations normalize the elevated intraocular pressure either by increasing the drainage of aqueous humor or decreasing the formation of aqueous. The drainage can be enhanced by following methods.

1. Opening the preexisting narrow angle of the anterior chamber.
2. Creating a subconjunctival filtering cicatrix.
3. Creating a communication between the anterior chamber and the suprachoroidal space.

Primary angle-closure glaucoma is ideally managed by laser iridotomy (Figs 27.11A and B) during the prodromal stage or stage of constant instability. The iridotomy prevents an attack of angle-closure glaucoma. It can also be performed once the acute attack of angle-closure glaucoma has been subsided by medication and goniosynechiae are not formed.



Figs 27.11A and B: Laser iridotomy

Laser Iridotomy

Laser iridotomy is indicated in following conditions:

1. Pupillary block glaucoma
2. Acute angle-closure glaucoma after breaking the attack medically
3. To prevent pupillary block in a high risk eye.

The presence of a complete synechial closure of the angle of the anterior chamber is a contraindication for laser iridotomy.

Laser iridotomy is performed using a condensing contact lens. 50 µm spot size and 800-1000 mW power are used in an argon laser for 0.02-0.10 second duration. Difficulty is experienced in very dark or very light irides. The Q-switched Nd:YAG laser is preferred in most eyes irrespective of color. It requires less power and less pulses to create an iridotomy.

Complications of laser iridotomy include focal lens opacity, corneal burn, acute rise in IOP and iritis.

Laser Gonioplasty or Peripheral Iridoplasty

Laser gonioplasty is indicated in plateau iris and synechial angle-closure of short duration. The angle that is closed by plateau iris will not open by laser iridotomy, therefore, laser gonioplasty is performed.

A spot size of 200-500 µm and 200-500 mW of power are used for 0.1-0.5 seconds. Stromal burns are created in the peripheral iris to cause contraction and flattening of the iris.

Incisional Surgery for PACG

Peripheral Iridectomy

When the laser iridotomy cannot be performed due to cloudy cornea, a flat anterior chamber or poor patient's cooperation, an incisional peripheral iridectomy (Fig. 27.12) is indicated.

The surgical technique of peripheral iridectomy is simple and safe. It is generally performed under local anesthesia. A self-sealing 1.5-2 mm corneal incision is made and the iris is grasped at the periphery with iris forceps. A small full-thickness piece of iris is excised. The iris is reposited with the iris repositor and the anterior chamber is reformed with air or Ringer's lactate.

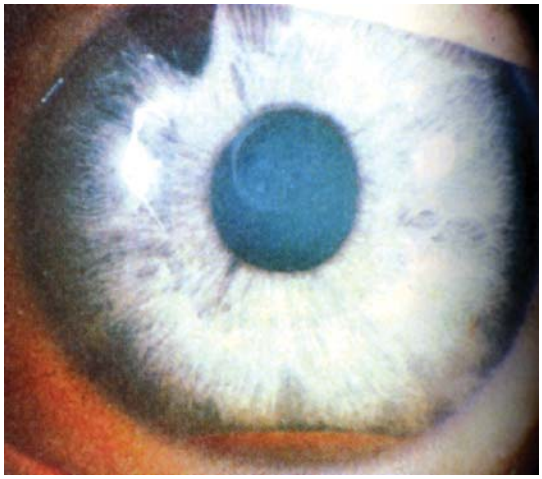


Fig. 27.12: Peripheral iridectomy

Basal Iridectomy

In the early chronic congestive phase of angle-closure glaucoma, the angle of the anterior chamber can be opened to permit adequate drainage of aqueous humor by performing a basal iridectomy. In this operation, the iris is torn from its ciliary attachment to obtain a broad opening at the periphery. The iridectomy may be a peripheral or a sectorial involving the sphincter pupillae.

Chamber Deepening and Goniosynechialysis

An anterior chamber deepening can be achieved by performing a paracentesis. A viscoelastic agent should be injected in the anterior chamber and a cyclodialysis spatula can be used to break the synechiae of recent onset.

Trabeculectomy

Trabeculectomy is indicated in chronic angle-closure glaucoma and when the peripheral iridectomy fails to normalize the IOP in patients with PACG.

Combined Glaucoma and Cataract Surgery

When cataract is associated with glaucoma, lens extraction should be considered in combination with trabeculectomy (video).

SURGERY FOR PRIMARY OPEN-ANGLE GLAUCOMA

Medical treatment is almost always employed as a primary modality for the management of primary open-angle glaucoma (POAG) and surgical intervention is only done when the medical regimen fails to control the intraocular pressure effectively. Glaucoma surgery is essentially aimed to reduce the intraocular pressure to a level at which progression of the disease is halted. It is not rare to find that despite the normalization of intraocular pressure some patients continue to lose vision and show progressive visual field defects and cupping of the disk.

Argon Laser Trabeculoplasty

Argon laser trabeculoplasty (ALT) is indicated in patients with POAG, pigmentry glaucoma and glaucoma associated with exfoliation syndrome.

The procedure of ALT consists of application of 50 evenly spaced laser burns at the junction of the anterior nonpigmented and posterior pigmented edge of the trabeculum (Fig. 27.13). ALT produces shrinkage of the treated area and stretching of trabecular meshwork. The shrinkage helps in opening the intratrabecular spaces and reverses the collapse of Schlemm's canal.

ALT causes a transient rise of IOP in about 20% of patients. Topical apraclonidine 1% or brimonidine 0.2% twice daily is recommended to control the postoperative pressure rise.

ALT is effective in 80% of cases of open-angle glaucoma with an average 8 mm Hg reduction of ocular tension. It is not a substitute for the medical therapy for glaucoma, but it can delay the surgical intervention. Besides argon laser, diode laser can also be used for trabeculoplasty.

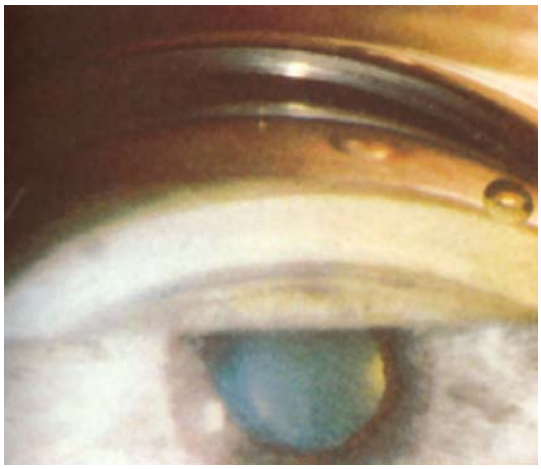


Fig. 27.13: Gonioscopic view of ALT

Filtration Operations

Filtering surgeries are fistualizing procedures which create a fistula for the flow of aqueous humor from the anterior chamber into the subconjunctival and sub-Tenon's space. Full-thickness filtering procedures such as sclerocorneal trephining, iridencleisis and thermal sclerostomy have fallen in disuse because of high complication rate.

Trabeculectomy

Trabeculectomy is a guarded partial-thickness filtering procedure described by Crains. Because of lower incidence of postoperative complications, it is the most preferred surgical procedure for the management of primary open-angle glaucoma.

Indications

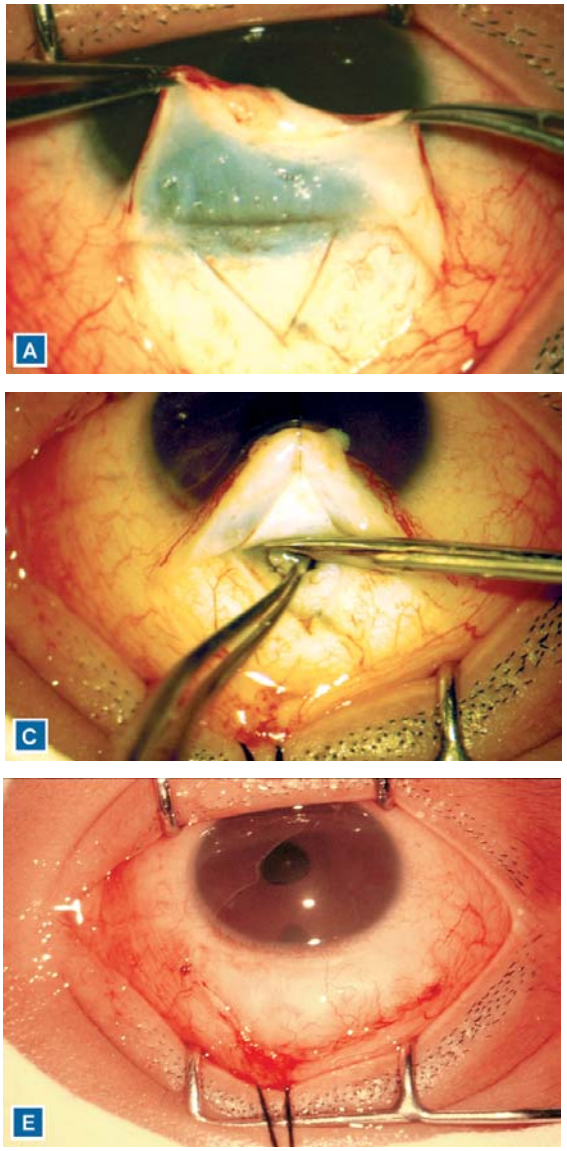
Indications for trabeculectomy include:

1. Maximal tolerable medical therapy when fails to reduce IOP to target level
2. The patient is unable to comply with the medical regimen
3. The medical therapy has to be discontinued owing to side effects
4. In spite of medical treatment, optic neuropathy and visual field defects progress.

Procedure

Trabeculectomy is performed under the following surgical steps (Figs 27.14A to E):

1. *Anesthesia*: Topical and infiltration anesthesia is preferred.
2. *Exposure:* The superior limbus is exposed by applying a corneal traction suture or superior rectus bridle suture.
3. *Conjunctival flap:* A fornix-based or limbal-based conjunctival flap is fashioned. The limbal-based flap allows a more secured closure. The incision is given 8 to 10 mm posterior to the limbus either superiorly at 12 o'clock or in superotemporal quadrant. Tenon's capsule is reflected and the underlying sclera is exposed.



Figs 27.14A to E: Steps of trabeculectomy: (A) Limbus-based conjunctival flap made and partial thickness triangular scleral flap marked, (B) Partial thickness scleral flap dissected, (C) Deep trabecular tissue excised, (D) Iridectomy performed, (E) Scleral and conjunctival incisions closed (*Courtesy*: Dr AK Mandal, LVPEI, Hyderabad)

4. *Scleral flap*: A 3-4 mm triangular limbal-based scleral flap is marked (Fig. 27.14A) and its two-third thickness is dissected anteriorly into the clear cornea (Fig. 27.14B).
5. *Paracentesis*: Paracentesis is performed for gradual lowering of IOP and maintenance of the anterior chamber.
6. *Trabeculectomy or excision of trabecular tissue:* A narrow strip of deeper sclera near the cornea containing the trabecular meshwork is excised (Fig. 27.14C) using corneal scissors or Kelly's punch.
7. *Iridectomy*: A peripheral iridectomy is performed (Fig. 27.14D) to lessen the risk of iris occluding the sclerostomy and preventing the pupillary block.
8. *Closure of the scleral flap:* The scleral flap is reposited and sutured tightly to avoid early shallowing of the chamber. Later, if required, laser suturolysis can be performed to promote filtration.
9. *Closure of the conjunctiva*: The flow of the aqueous should be tested around the flap before the conjunctiva is closed. The conjunctival flap is reposited. The fornix-based flap is sutured by two interrupted sutures at limbus while the limbus-based by continuous sutures (Fig. 27.14E).
10. *Patching of eye*: After a subconjunctival injection of antibiotic-corticosteroid, the eye is patched.

In trabeculectomy, the aqueous is drained underneath the lamellar flap of sclera and overlying conjunctiva, and also to some extent through the cut end of Schlemm's canal. It gives a flat well-protected bleb (Fig. 27.15).

Antifibrotic Agents

The use of antifibrotic agents such as mitomycin-C (MMC) and 5-fluorouracil (5-FU) during trabeculectomy results in greater success in lowering IOP. The use of antifibrotic agent is advocated for high risk patients with glaucoma. It is indicated in following conditions:

1. Aphakic or pseudophakic glaucoma
2. Neovascular glaucoma, and
3. Patients with failed filtering surgery.

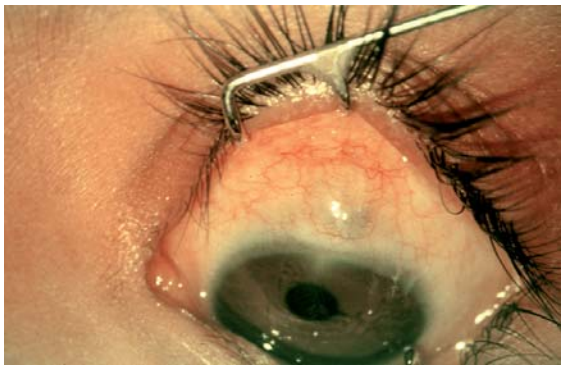


Fig. 27.15: Well-formed bleb after filtration operation (*Courtesy*: Dr AK Mandal, LVPEI, Hyderabad)

Complications

Complications of filtering surgery may occur either early (within 3 months of surgery) or late.

Early complications include hyphema, uveitis, shallow or flat anterior chamber, cystoid macular edema and hypotony.

Late complications include bleb failure with rise of IOP, bleb leakage, over-filtering bleb (Fig. 27.16), hypotony maculopathy, cataract and bleb-related endophthalmitis. The risk of endophthalmitis is greater after the use of antifibrotic agent.

Nonpenetrating Glaucoma Surgery

Nonpenetrating glaucoma surgery lowers the IOP and avoids some of the complications of trabeculectomy. A deep sclerectomy under a scleral flap without entering the anterior chamber may be performed with or without a collagen implant in nonpenetrating glaucoma surgery.

Fig. 27.16: Over-filtering bleb

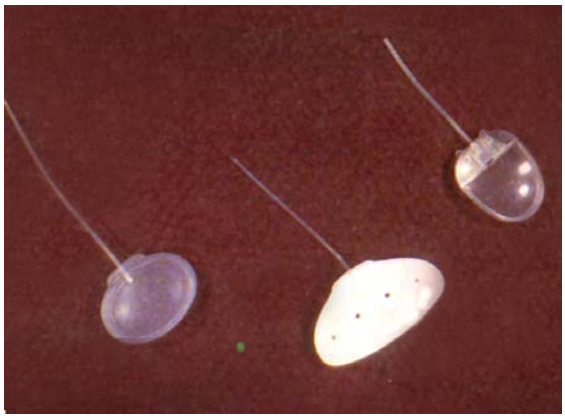


Fig. 27.17: Tube shunts from left to right: Krupin disk, Baerveldt and Ahmed valve (*Courtesy*: Dr TW Perkins, Madison)

Viscocanalostomy is a procedure wherein a deep sclerectomy is performed and viscoelastic material is injected into Schlemm's canal to expand it.

SURGERY FOR SECONDARY GLAUCOMAS

Secondary glaucomas have a varied etiology. The choice of surgery largely depends on the pathogenesis of glaucoma, for example, phacomorphic glaucoma is managed by extraction of the lens, angle recession glaucoma by trabeculectomy and neovascular glaucoma by setons or cycloablative procedures.

Setons

Setons are silicone/silastic narrow tubes to shunt the aqueous from the anterior chamber to the subconjunctival space. Molteno's, Ahmed's, Shocket's and Krupin-Denver valve implants (Fig. 27.17) are currently in use to control the IOP in neovascular (Fig. 27.18) or aphakic glaucoma nonresponsive to standard filtration techniques.

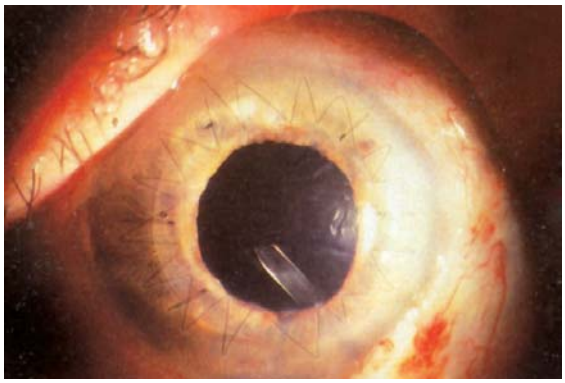


Fig. 27.18: An inferiorly placed tube shunt implant in a patient wih neovascular glaucoma following PKP (*Courtesy*: Dr TW Perkins, Madison)

Cycloablative Procedures

Cyclodialysis

Cyclodialysis is an internal bypass surgery in which disinsertion of the ciliary body from its scleral attachment forms a communication between the anterior chamber and suprachoroidal space. This lowers the IOP effectively by outflow of aqueous humor through suprachoroidal space and by less production of aqueous by the detached ciliary body. The operation is indicated in aphakic glaucoma.

A conjunctival flap is made in the upper and outer quadrant of the globe. A 3 mm long radial incision is made at the limbus. The cyclodialysis spatula is gently inserted between the sclera and the ciliary body. It is gradually pushed towards the cornea until its tip appears in the anterior chamber. The spatula is swept on either side to

separate the ciliary body from the scleral spur. It is then withdrawn and the conjunctival flap is sutured. Hyphema is a common complication which can be prevented by injection of air in the anterior chamber.

Cyclodiathermy

The formation of aqueous humor can be reduced by inducing segmental atrophy of the ciliary body either by application of diathermy current or by cryopexy. Cyclodiathermy may be either a surface type or a penetrating type.

Surface diathermy can be applied over the conjunctiva. A single row of applications are made 3 mm behind the limbus. An electrode of 2 mm diameter is used and a current of 50 to 60 milliamperes is passed at each point for about 15 seconds.

Penetrating diathermy is applied after reflection of the conjunctiva. Nearly 60 to 80 punctures are made with a needle 0.5 mm long and 0.2 mm thick in a zone 3 mm behind the limbus. A current of 50 to 60 milliamperes is used for one second.

A partial destruction of ciliary body by diathermy may occasionally lead to an irreversible ocular hypotonia due to atrophy of the ciliary epithelium. It is, therefore, advisable to monitor the procedure in different sittings; the surface diathermy is preferred over the penetrating.

Cyclocryopexy

Cyclocryopexy is relatively a safe alternative to induce segmental atrophy of the ciliary body. In this procedure transscleral freezing of the ciliary body is accomplished by applications of a freezing probe over a segment of the ciliary body (Fig. 27.19). Ordinarily, the freezing of the tissue at –70°C for a period of 45 seconds gives the desired effect. It is relatively a less destructive procedure than cyclodiathermy.

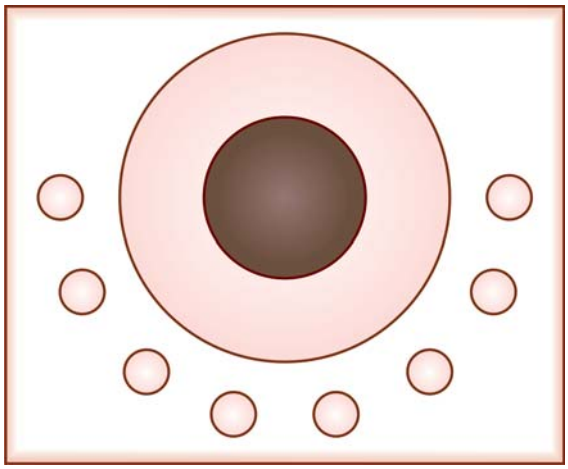


Fig. 27.19: Cyclocryopexy

Cyclophotocoagulation

Transscleral cyclophotocoagulation is an effective method of lowering the IOP in advanced cases of glaucoma. It destroys the areas of nonpigmented epithelial cells of ciliary body resulting in decreased aqueous production. The procedure includes 30 to 40 applications of 4 to 6 Joules energy setting of 20 ms durations, using Nd: YAG laser, 3 mm away from the limbus 360° over the pars plicata.

SURGERY FOR CONGENITAL CATARACT

Congenital cataract causing serious visual impairment needs surgery. It is advisable to operate upon congenital cataracts at any time after the infant is six weeks of age. The prognosis is good in patients with bilateral cataracts unassociated with nystagmus. Unilateral congenital cataract has relatively poor prognosis.

Several operative techniques for the management of congenital cataract such as: (i) optical iridectomy, (ii) discission, (iii) linear extraction

(iv) aspiration, (v) phacoaspiration, and (iv) pars plana lensectomy, are available. Because of the inevitable danger of vitreous loss, intracapsular lens extraction is disfavored.

Currently optical iridectomy, discission and linear extraction are not performed owing to the availability of better surgical procedures. Phacoaspiration and pars plana lensectomy are often preferred. In children above the age of 2 years, intraocular lens (IOL) implantation with primary posterior capsulotomy and anterior vitrectomy are recommended for the prevention of posterior capsular opacification.

Aspiration

Aspiration is performed under general anesthesia in infants and children. The pupillary dilatation must be maximal. A self-sealing corneal incision is made. Anterior capsulotomy is done under a viscoelastic agent using a cystotome or a bent 26 G needle. The lens matter is aspirated using an irrigation-aspiration canula (Simcoe canula) or the irrigation-aspiration handpieces of a phacoemulsification machine. The anterior chamber is reformed, antibiotic and cycloplegic are applied and eye is patched.

Lensectomy

Lensectomy is indicated in traumatic cataract, complicated cataract, subluxated lenses and congenital cataract. The operation can be performed through the pars plana or the pars plicata or the limbus. A vitreous cutter is used with a second port for infusion. The posterior capsule can be left behind for secondary IOL implantation. In the posterior route the whole lens, including anterior and posterior capsules, is removed. In complicated cases, the pupillary aperture is enlarged by severing the synechiae and, if needed, anterior vitrectomy can be performed to prevent the development of postoperative inflammatory membrane and secondary glaucoma.

Complications

In congenital cataract, a good surgical result does not necessarily mean a good vision. Amblyopia, secondary cataract, iridocyclitis, secondary glaucoma and retinal detachment may occur after the surgery.

Correction of Pediatric Aphakia

The risk of visual deprivation amblyopia has an impact on the timing of surgery as well as correction of aphakia. Prompt correction of postoperative aphakia is necessary to prevent amblyopia. Refraction becomes nearly stable after one week of surgery due to small size of incision. Aphakia can be corrected by following methods.

1. *Spectacle correction*: Children older than one year with bilateral aphakia may tolerate the aphakic spectacles well.
2. *Contact lenses*: Silicone soft contact lenses are used for extended wear for the correction of monocular or binocular aphakia. Parents can fit and remove the lenses on a weekly basis.
3. *IOL implantation*: Bilateral intraocular lens implantation in the posterior chamber during cataract surgery is an established approach to correct pediatric aphakia. It provides good visual acuity and binocularity. Children with aphakia may be corrected by secondary lens implantation.

SURGERY FOR SENILE CATARACT

Senile cataract extraction is by far the most common operation performed by ophthalmologists all over the world. The introduction of fine needles, suture material and operating microscope has made the cataract surgery very safe. The phacoemulsification technique and small incision cataract surgery (SICS) have improved the quality of vision and reduced the convalescence time of the patient.

Preoperative Evaluation

The following general and ocular evaluations of the patient with cataract should be carried out before undertaking the surgery.

General health: All cases of cataract must be examined in consultation with a physician to exclude diabetes, hypertension, ischemic heart disease, chronic obstructive pulmonary disease, urinary obstruction and bleeding disorders. These diseases do not preclude cataract extraction but must be controlled before the surgery. Information about medication and drug sensitivities that might alter the result of surgery should be obtained. Physical conditions like obesity, kyphosis and head tremors may need a change in the surgical approach.

Ocular history: A positive history of ocular trauma, inflammation, amblyopia, retinal disorders and glaucoma is likely to alter the visual prognosis. If patient has had cataract surgery in the fellow eye or refractive surgery, past records should be reviewed along with the information about any operative or postoperative complications.

Visual Functions

Visual acuity: The near and the distant visual acuity is tested and refraction is performed to determine the best corrected visual acuity. Contrast sensitivity may be recorded in patients with cataract even when Snellen's acuity is good. The patient with a mature or a hypermature cataract should have at least perception of light (PL) vision.

Visual field and projection of rays: Confrontation visual field testing documents visual field loss in patients with immature cataract, but in dense cataract projection of rays may be helpful to test the peripheral visual field.

Potential acuity: Estimation of potential acuity helps in accessing the visual loss due to cataract formation. Laser interferometer and potential acuity meter may be used for this purpose. A monochromatic laser light forms a diffraction fringe pattern on the retina independent of the lens opacities in laser interferometry. The visual acuity may be measured by varying the spacing of the fringe, however, it is not accurate in detecting small foveal lesions especially in dense cataract. The potential acuity meter measures the visual acuity by projecting a Snellen's chart or numericals through the pupil. The images can be projected around the lenticular opacities.

Macular Function Tests

1. *Two pinholes test*: Macular function is roughly tested by asking the patient to look through an opaque disk perforated by two closely placed pinholes behind which a light is held. If the patient can perceive two lights, the macula is probably functioning normally.
2. *Maddox rod test*: The patient is asked to look at a distant light through a maddox rod. The perception of a continuous and unbroken red line suggests that the macula is probably normal.
3. *Entoptic view test*: When a bare lighted bulb of an ophthalmoscope is rubbed on the closed lower eyelid, an intelligent patient can perceive the vasculature of retina and may appreciate and describe scotoma in the field. Entoptic viewing of white blood cells of perifoveal capillaries may be appreciated on an intensely illuminated blue-light background (blue-light entoptoscopy).
4. *Color vision*: The patient's ability to identify different colored lights projected from an ophthalmoscope provides an indication of macular function.

Slit-lamp Examination

In order to exclude the presence of an active intraocular inflammation, slit-lamp examination must be carried out. The presence of cells in the aqueous humor is a contraindication for any

intraocular surgery. Slit-lamp helps in detailed evaluation of anterior segment of the eye.

Ultrasonography

Ultrasonography, especially the B-scan, produces a two dimensional picture of the vitreous and the retina thus gives valuable information about the posterior segment of the eye in presence of a dense cataract precluding funduscopy.

Electrophysiological Tests

When the results of other tests are inconclusive electroretinogram and visual evoked potential tests should be done to evaluate the status of the retina and the optic nerve, respectively.

Measurements

Before undertaking cataract surgery, following measurements should be performed :

1. *Refraction*: Refraction, if possible, should be performed in both the eyes to achieve post-operative emmetropia or mono vision for correction of presbyopia.
2. *Biometry*: A-scan ultrasonography is necessary to calculate an appropriate IOL power for implantation.
3. *Corneal topography*: If the patient has already undergone refractive surgery, corneal topography provides a more accurate information about the corneal astigmatism than manual keratometry.
4. *Corneal pachymetry*: The measurement of corneal thickness provides an estimate of corneal endothelial function. When the thickness is more than 640 µm, the cornea is edematous and possibility of postoperative decreased corneal clarity is greater.
5. *Specular microscopy*: It is used to determine the endothelial cell count. Normally the cell count is more than 2400 cells/mm^2. A preoperative low cell count or loss of cells during cataract surgery is likely to result in postoperative corneal decompensation.
6. *Applanation tonometry*: Preoperative measurement of IOP is mandatory not only to exclude glaucoma but also to prevent intraoperative and postoperative complications.

Informed Consent

The risks and benefits of cataract surgery should be explained to the patient and the relatives, and a written informed consent should be obtained.

Preoperative Preparation

Preoperative lacrimal sac syringing and conjunctival swab culture are advised as routine investigations. Prophylactic broad-spectrum antibiotic like gatifloxacin (0.3%), moxifloxacin (0.5%), ciprofloxacin (0.3%) or tobramycin (0.3%) drops are instilled 4 times a day for 4 to 7 days prior to surgery.

Hypotonia must be obtained before the cataract surgery. In routine, two tablets of acetazolamide (250 mg) are given a night before operation as well as two hours before the operation. Most surgeons prefer to reduce intraocular pressure by manual massage or by applying pinkie ball for 20 minutes following peribulbar anesthesia. Patients of glaucomatous cataract should be given hyperosmotic agents like 20% mannitol intravenously an hour before the surgery.

The eye to be operated must be marked by an adhesive tape. Eyelashes are usually trimmed a day before surgery and the skin of the eyelids, lid margins and skin around the eye is cleaned with 10% povidone-iodine solution. Some surgeons use 5% povidone in the eye also.

The pupillary reaction should be brisk and the pupil should dilate readily with mild mydriatics. Topical NSAIDs like flurbiprofen (0.03%) or diclofenac sodium (0.1%) drops are instilled along with tropicamide (1%) and phenylephrine (5-10%) every 30 minutes for 2 hours before the surgery to obtain adequate and sustained pupillary dilatation intraoperatively.

Types of Cataract Extraction

Broadly speaking, two types of cataract extraction, intracapsular and extracapsular, are performed.

Intracapsular Cataract Extraction

The cataractous lens is removed with its capsule after rupturing the suspensory ligament of the lens (zonule) in intracapsular cataract extraction (ICCE). The technique of ICCE is quite simple.

Indications for ICCE

1. Subluxated and dislocated lenses
2. Cataract in those eyes with risk of development of phacoanaphylactic reaction
3. Surgeon is not trained in microsurgery or facilities for microsurgery are not available.

Contraindications for ICCE

1. Cataract in children and young adults
2. Traumatic cataract with ruptured capsule
3. High myopia, Marfan syndrome and vitreous presenting in the anterior chamber are relative contraindications.

Advantages of ICCE

1. The technique is simple and quick and does not need sophisticated instruments
2. The learning curve of ICCE is small
3. There is no chance of posterior capsular opacification as the capsule is removed with the lens, thus additional surgery is not required.

In spite of above advantages, ICCE is not a safe procedure mainly due to the large size of incision.

Disadvantages of ICCE

1. Wound healing is delayed
2. Complications like wound leak, vitreous incarceration, endothelial cell loss and cystoid macular edema are more common after ICCE than after extracapsular cataract extraction.
3. Postoperative astigmatism is significant
4. Posterior chamber IOL implantation becomes problematic after ICCE
5. Visual rehabilitation is significantly delayed.

Extracapsular Cataract Extraction

In extracapsular cataract extraction (ECCE) the lens is removed leaving behind a peripheral part of its anterior capsule as well as the entire posterior capsule. The technique is safe and most suited for posterior chamber intraocular lens (PCIOL) implantation. Ideally all cases of cataract should be operated by extracapsular technique unless otherwise contraindicated. Currently, planned ECCE with IOL implantation is the most preferred surgery.

Indications for ECCE

1. All patients in whom posterior chamber IOL implantation is planned
2. Cataract in young adults with capsulohyaloidal adhesions and strong zonule
3. Cataract associated with high myopia and degenerated vitreous
4. Cataract with fellow eye having aphakic retinal detachment following ICCE
5. Complicated cataract.

Contraindications for ECCE

Practically there is no contraindication for ECCE. However, it should be done meticulously in patients with risk of development of phacoanaphylactic reaction.

Advantages of ECCE

1. It is a safe procedure and rate of complication is very low
2. ECCE can be performed in patients of all ages

3. It is a most suited procedure for posterior chamber lens implantation
4. Visual recovery is early and very good due to relatively low astigmatism.

Disadvantages of ECCE

1. It is relatively more time consuming operative technique than ICCE
2. ECCE requires sophisticated instruments and an operating microscope
3. The surgeon needs training for ECCE and its learning curve is comparatively steeper than ICCE
4. Posterior capsular opacification is the main drawback of ECCE which needs Nd:YAG laser capsulotomy.

Intracapsular Cataract Extraction

Intracapsular cataract extraction is performed in following steps.

Anesthesia and Akinesia

Surface and regional anesthesia and akinesia are obtained by topical instillation of 4 percent xylocaine or 0.5% proparacaine drop 3 to 4 times and infiltration of 7 ml of anesthetic solution in the peribulbar space. The solution contains 2% lignocaine/xylocaine and 0.5% bupivacaine (in a ratio of 2:1 volume), 150 IU hyaluronidase and adrenaline (1:200000). Of this solution, 3.5 ml is injected at the inferior orbital rim 1 cm medial to the lateral canthus and the remaining just above the supraorbital notch. It is followed by application of pad and pinkie ball (pressure of about 30 mm Hg) or intermittent digital massage for 20 minutes. This method obviates the need for facial block and produces a soft eye. After removal of pinkie ball, the skin of the eyelids and around the eye is again cleaned with 10% povidone-iodine solution.

Retraction of Eyelids

The lid is retracted by application of an eye speculum or sutures. If an eye speculum is used for the separation of the lids it should not press upon the globe.

Fixation of Eyeball

The superior rectus suture is passed to fix the eye in the downward gaze.

Preparation of Conjunctival Flap

The operation can be performed without fashioning a conjunctival flap but the flap protects the corneoscleral section and helps in the healing of the wound. A fornix-based conjunctival flap is preferred as it practically eliminates the chance of epithelial invasion of the anterior chamber.

Hemostasis

Fashioning of a conjunctival flap causes bleeding which can be checked by wet-field cautery or heat cautery.

Limbal Groove and Preplaced Sutures

The application of 3 preplaced sutures helps the surgeon to close the wound in the event of adversities. Fine nonabsorbable suture (prolene or polyamide 10-0) may be used as the suturing material. The sutures are generally placed after making a groove or gutter at the limbus.

Sclerocorneal or Corneal Section

The sclerocorneal section (Fig. 27.20A) can be made either with the Bard-Parker knife or a blade and corneal scissors. The anterior chamber is reached by deepening the limbal groove. The incision is enlarged from 3 o'clock to 9 o'clock position with the corneal scissors.

Extraction of the Lens or Lens Delivery

The intracapsular lens extraction can be performed by various techniques. The lens can be removed by the intracapsular lens forceps or cryoprobe.

Forceps delivery: The capsule forceps is introduced closed in the anterior chamber and moved over the anterior capsule of the lens to reach its thickest part at 6 o'clock position. Then the limbs are opened 2 mm, pressed gently backwards to engage the capsule and closed. The lens is slightly lifted and suspensory ligaments are ruptured by making zig-zag movements. The lens tumbles forwards. The lower part of the lens is supported by a lens expressor. The lens is coaxed out of the wound by continuing gentle pressure by the expressor.

The lens can also be delivered by catching its upper pole and breaking the zonule with the help of a lens expressor. Once the zonular attachment is severed, it is slided out of the wound.

Smith's tumbling technique: The hypermature milky-white cataractous lens can be tumbled out by applying pressure at 6 o'clock and counter-pressure at 12 o'clock positions (Smith's technique). The technique requires practice otherwise vitreous loss may occur.

Cryoextraction: Cryoextraction is preferred for the intracapsular lens removal (Fig. 27.20B) over other techniques. A temperature of –35° to –40° C creates a good adhesion between the probe and the lens. The cryoprobe is applied on the mid-periphery of the superior pole of the lens after retracting the iris and lifting the cornea. The lens is removed through the wound by gently elevating it and moving it from side-to-side to break the zonular attachments. Care must be taken that the probe should not come in contact with the iris or the cornea. If the iris sticks, the probe can be freed by irrigation with Ringer's lactate.

Zonulysis: The intracapsular lens extraction in young adults may present difficulty owing to toughness of the zonule. It can be overcome by the use of zonulysin (zonulotome, α-chymotrypsin), a proteolytic enzyme which dissolves the zonule. Injection of a small quantity of 1 in 5000 solution of zonulysin into the posterior chamber results in fragmentation of the zonule within 2 to 3 minutes. The excess of the enzyme is thoroughly irrigated out. Thereafter, the lens can be easily expressed out with a gentle pressure by a lens expressor or may be removed by a cryoprobe. Nowadays, this surgery has become obsolete and extracapsular cataract extraction or phacoemulsification has replaced it.

Iridectomy

A button-hole peripheral iridectomy is almost always performed in intracapsular cataract surgery. The purpose of an iridectomy is to create a communication between the posterior and anterior chambers of the eye, to help in the reformation of the anterior chamber and to avoid a possible postoperative prolapse of the iris into the wound. Occasionally, a sector iridectomy is necessitated in rigid contracted pupil to facilitate the lens delivery. For sector iridectomy, first a peripheral iridectomy is done and then the tip of the 'V' is cut to the pupillary border of the iris using a Vanna's scissors.

Reformation of Anterior Chamber

After the iridectomy, the iris is reposited into the anterior chamber. The chamber is reformed by injection of an air bubble or irrigating fluid like Ringer's lactate or balanced salt solution (BSS) into the anterior chamber.

In case pupil is widely dilated, pupillary constriction can be obtained by the use of intracameral pilocarpine (0.5%).

Anterior Chamber Lens Implantation

An anterior chamber IOL can be inserted after the pupillary constriction (Fig. 27.20C) and filling the anterior chamber with a viscoelastic agent. The

inferior haptic of the lens is inserted into the anterior chamber angle and the superior haptic into the superior chamber angle with the help of a lens holding forceps. The viscoelastic is removed and is replaced with BSS.

Wound Closure

The preplaced sutures are tied and additional end-to-end postplaced sutures are applied to ensure proper apposition of the wound. Some surgeons prefer to remove the preplaced sutures and close the section by continuous running sutures.The fornix-based conjunctival flap is pulled over the section and retained there by conjunctival sutures or coaptation using a wet-field cautery, if required.

To prevent postoperative infection and inflammation, 10 mg/0.5 ml gentamicin and 0.5 mg/0.5 ml dexamethasone are injected subconjunctivally. The operated eye is patched with a sterile pad and an adhesive tape and/or a bandage.

Conjunctival stitches are removed on the 7th day and buried sclerocorneal sutures need not be removed. Generally, glasses are prescribed after 6- 8 weeks of the operation.

Extracapsular Cataract Extraction

The operation is performed in following steps.

The initial surgical steps (anesthesia, lid retraction, superior rectus bridling, raising of conjunctival flap and cauterization) of ECCE are similar to that of ICCE.

Limbal Incision

The limbal groove is made from 2 to 10 o'clock and the anterior chamber is entered by a stab incision with a blade at 12 o'clock position.The anterior chamber is reformed by injecting a viscoelastic material.

Anterior Capsulotomy

Anterior capsulotomy is performed either by a cystotome (Fig. 27.20D) or a bent 26 gauge needle in a maintained anterior chamber to minimize the corneal endothelial damage.

Depending upon the shape, the capsulotomy techniques are named as can-opener, envelope and continuous circular capsulotomy. The continuous circular capsulotomy or *capsulorhexis* is the most desired procedure especially for phacoemulsification with posterior chamber IOL implantation.

The cut piece of anterior capsule is removed with the help of a McPherson forceps.The corneoscleral section is completed from 2 to 10 o'clock position. Since only the nucleus of the lens has to be delivered, the section is smaller than that required for ICCE.

Hydrodissection

The next step is hydrodissection. The irrigating solution is slowly injected in different quadrants underneath the edge of anterior capsule to separate the cortex and nucleus from the posterior capsule.

Nucleus Removal

The nucleus is removed by depression of the posterior lip of the section with a vectis and counter pressure with a lens expressor at 6 o'clock (Fig. 27.20E).

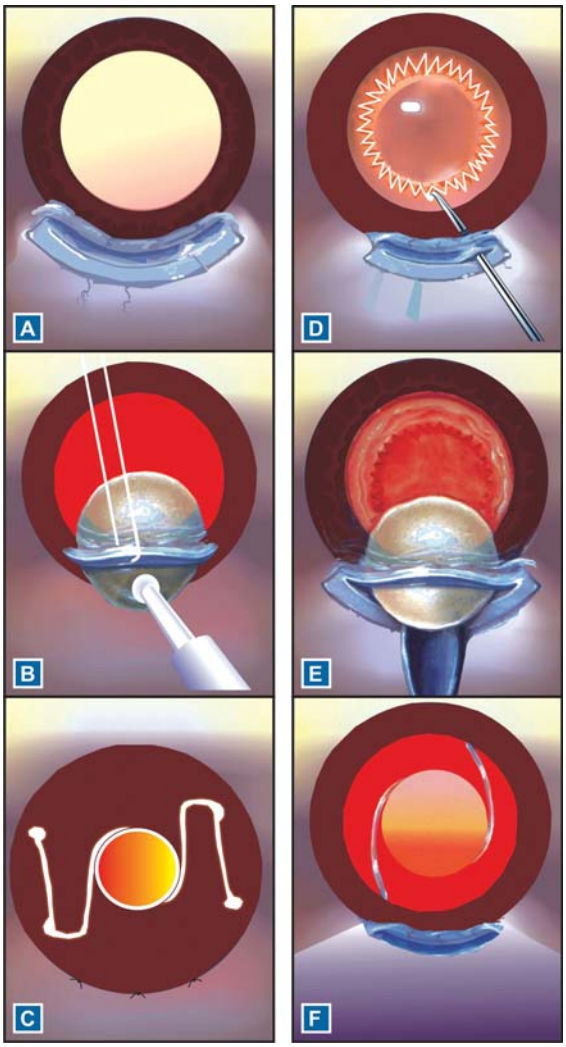
Cortical Aspiration

Aspiration of the cortex is performed using a Simcoe canula. The cortex should be picked up carefully without capturing the posterior capsule.

IOL Implantation

After aspiration of the cortex, the anterior chamber is reformed with a viscoelastic substance and a posterior chamber IOL is implanted in the capsular bag (Fig. 27.20F).

If only planned ECCE has to be done, one may perform a peripheral iridectomy, reposit the iris, reform the anterior chamber with BSS, close the wound and inject an antibiotic-corticosteroid solution subconjunctivally (identical steps of ICCE). In place of a miotic, a mydriatic may be instilled and the eye is patched.



Figs 27.20A to F: ICCE: (A) Large limbal incision, (B) Cryoextraction of the lens, (C) An anterior chamber IOL is implanted and wound is closed with interrupted sutures ECCE: (D) A can-opener capsulotomy is performed, (E) Nucleus is being removed by vectis, (F) A posterior chamber IOL is implanted

Postoperative Care

Postoperatively an aggressive course of topical corticosteriods should be administered to reduce the inflammatory reaction. Topical corticosteroid drops are instilled 8 times a day in the first week, followed by 6 times a day in the second week, 4 times a day during the third week and 3 times a day in the fourth week. Cycloplegic drop once daily is advised for one month. Postoperatively IOP may rise, therefore, IOP must be checked on each follow-up visit.

Small Incision Cataract Surgery

Small incision cataract surgery (SICS) is a type of extracapsular cataract extraction. It is a safe and sutureless surgery which can be performed even in a hard brown cataract with ease. The steps of surgery are similar to ECCE with some modifications in the external scleral incision and delivery of the nucleus.

A straight or frown-shaped, 5.5-6.5 mm incision is made nearly 2 mm behind the limbus (Fig. 27.21) and a scleral tunnel is prepared. To make the incision self-sealing (sutureless), a pocket is created along the curvature of the limbus (Fig. 27.22). The internal incision of the tunnel is larger than the external to accommodate the lens.

A side-port entry is made that helps in filling the anterior chamber with a viscoelastic agent

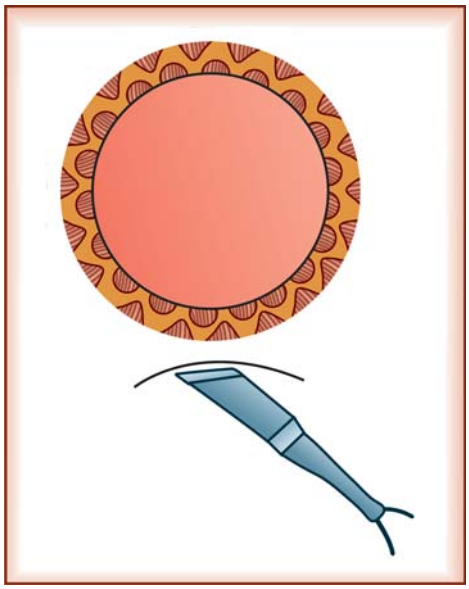


Fig. 27.21: Frown scleral incision

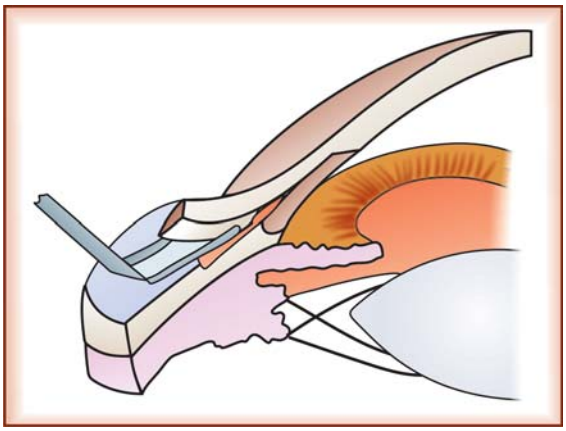


Fig. 27.22: A scleral tunnel is dissected

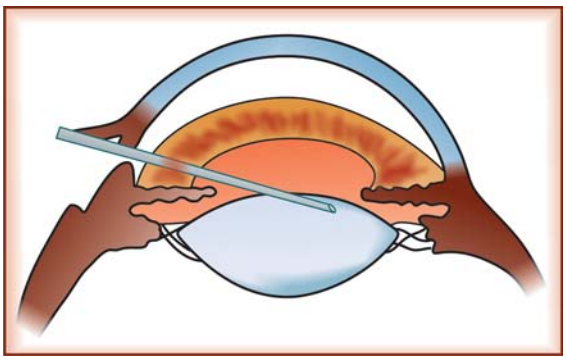


Fig. 27.23: Hydrodissection

before anterior capsulotomy. Depending on the size of the nucleus, a capsulorhexis is performed.

Multiquadrant hydrodissection (Fig. 27.23) facilitates the prolapse of the nucleus in the anterior chamber by a lens hook (Fig. 27.24) or one can use a bent 26 G needle to bring the nucleus out of the capsular bag. An irrigating vectis is used for the delivery of the nucleus or it can be removed by modified Blumenthal technique (Figs 27.25A and B). The cortical cleaning is achieved by a gentle flushing with viscoelastic and irrigation-aspiration using a Simcoe canula. Care must be taken during subincisional cortical aspiration.

The capsular bag is inflated with viscoelastic and IOL is implanted in-the-bag. The viscoelastic is removed and the anterior chamber is reformed with BSS.

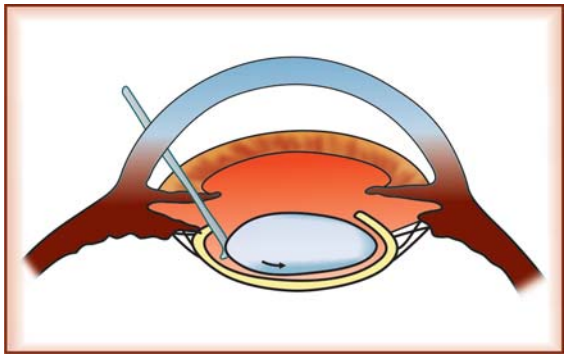
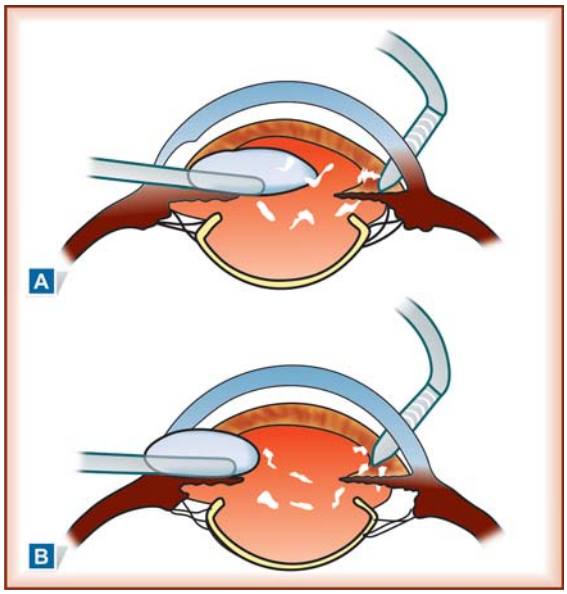


Fig. 27.24: Nucleus is being prolapsed out of the capsular bag



Figs 27.25A and B: Removal of the lens by modified Blumenthal technique

(Figs 27.21 to 27.25 are reproduced by the *courtesy* of Drs KPS Malik and Ruchi Goyal from their book, Small Incision Cataract Surgery, published by CBS Publishers, New Delhi)

Phacoemulsification

Phacoemulsification is essentially an extracapsular lens extraction performed with the help of a phacoemulsification machine. The phacoemulsifier provides controlled irrigation-aspiration and

phacoemulsification. The phacoemulsification is performed by a phaco handpiece the tip of which vibrates 28000-60000 cycles per second. The emulsified lens is removed by aspiration-infusion.

The technique has following steps (video).

1. *Side-port entry:* A side-port entry is made for filling the anterior chamber with viscoelastic substance (Fig. 27.26).
2. *Clear corneal incision:* A 3-3.5 mm incision in the superotemporal clear cornea with a diamond keratome (Fig. 27.27) or a scleral tunnel incision with an internal corneal lip is made. It is a self-sealing incision and does not require stitching after the surgery.
3. *Continuous curvilinear capsulorhexis (CCC):* The anterior chamber is filled with viscoelastic and a 6 mm capsulorhexis is performed with the help of a bent 26 G needle (Fig. 27.28) or a Utrata forceps.
4. *Hydrodissection*: A small amount of BSS is injected between the anterior capsular rim and the cortex of the lens at 3-4 places for separation of peripheral cortex from the capsule.
5. *Hydrodelineation*: To obtain the separation of firm nucleus from the epinucleus, a small quantity of BSS is injected into the substance of the nucleus.
6. *Nuclear emulsification*: It is done by ultrasonic power of phaco handpiece either in the capsular bag or in the iris plane. In the *divide and conquer technique* a deep linear groove is sculpted in the nucleus. After the rotation of the nucleus to 90 degrees a further trenching at right angle to the previous groove is performed. Then the nucleus is divided into 4 quadrants with the help of a chopper and phaco tip.

In *direct chop technique* the nucleus is divided into two heminuclei using the phacoemulsification handpiece and a chopper. The heminuclei are then further subdivided into small pieces.

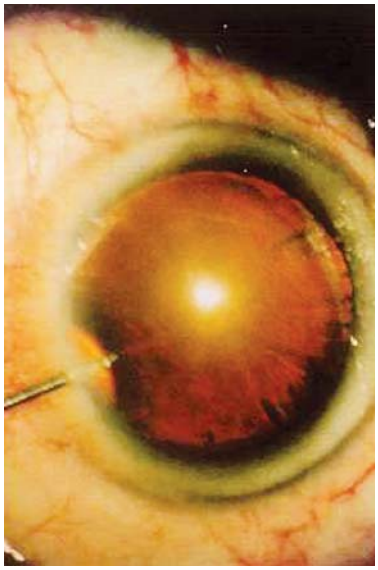


Fig. 27.26: Side-port entry (*Courtesy*: Dr Amar Agarwal, Chennai)

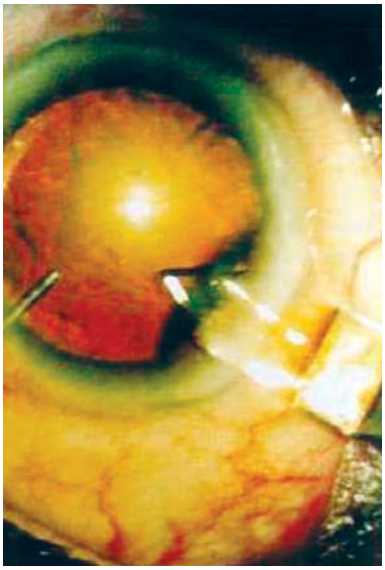


Fig. 27.27: Clear corneal incision (*Courtesy*: Dr Amar Agarwal, Chennai)

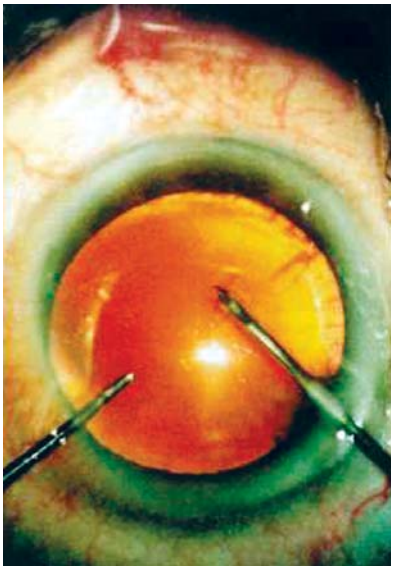


Fig. 27.28: Capsulorhexis with needle (*Courtesy*: Dr Amar Agarwal, Chennai)

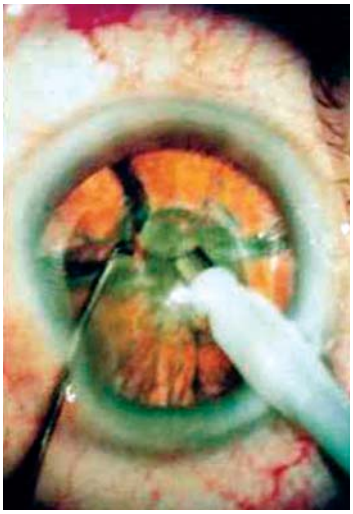


Fig. 27.29: Nuclear emulsification (*Courtesy*: Dr. Amar Agarwal, Chennai)

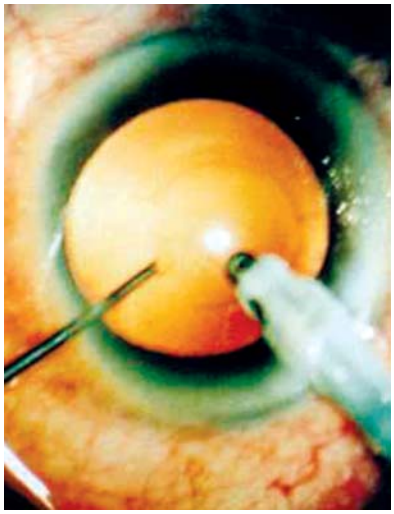


Fig. 27.30: Cortical cleaning (*Courtesy*: Dr. Amar Agarwal, Chennai)

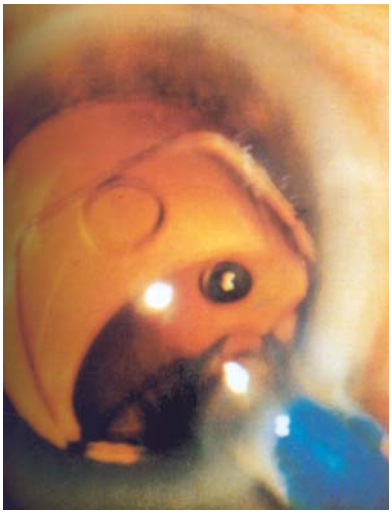


Fig. 27.31: Implantation of a plate haptic foldable lens (*Courtesy*: Dr. Amar Agarwal, Chennai)

7. *Quadrant removal*: It is performed with the help of the phaco probe. One-by-one each quadrant is emulsified and aspirated (Fig. 27.29). Then the epinucleus is aspirated.
8. *Cortical cleaning*: The remaining lens cortex is aspirated with the help of a coaxial or bimanual irrigation-aspiration canula (Fig. 27.30).
9. *Lens implantation*: A posterior chamber foldable lens can be implanted without enlarging the corneal incision (Fig. 27.31).

Advantages of Phacoemulsification

Phacoemulsification is currently the most popular surgical procedure for the removal of cataract. It has several advantages (Table 27.1).

Limitations of Phacoemulsification

Phacoemulsification has some limitations. They include high cost of the phaco machine, difficult technique requiring training under an expert, relatively long learning curve, and possibility of complications in hard cataract and compromised cornea.

Table 27.1: Advantages of phacoemulsification

1. Small incision, sutureless surgery
2. Relatively safe surgery
3. Maintains the anterior chamber throughout the surgery
4. Minimum postoperative astigmatism
5. Rapid convalescence

Complications of Cataract Surgery

Intraoperative Complications

The common intraoperative complications of cataract surgery are summarized in Table 27.2.

Retrobulbar hemorrhage may develop following the retrobulbar or peribulbar injection for regional anesthesia. Eye becomes proptosed and suffused. It usually takes two weeks for the hemorrhage to resolve.

Descemet's membrane detachment or stripping near the incision may occur due to instrumental injury.

Hyphema appears during the surgery owing to oozing from the corneoscleral wound or from the traumatized iris. The oozing points on the sclera

Table 27.2: Intraoperative complications of cataract surgery

1. Retrobulbar hemorrhage
2. Descemet's membrane detachment
3. Hyphema
4. Vitreous prolapse or loss
5. Lens subluxation or dislocation

should be cauterized and blood from the anterior chamber should be soaked out by a piece of surgical sponge or the anterior chamber be irrigated. Rarely, removal of clot with a forceps becomes necessary.

Rupture of the lens capsule during forceps delivery is not unusual especially in hypermature cataract during ICCE.

Vitreous presentation in the anterior chamber even before the extraction of the lens may occur in a traumatized eye or due to previous poorly performed intraocular surgery. *Prolapse of the vitreous* after the nuclear delivery can happen because of posterior capsular rent that occurs most commonly during cortical irrigation-aspiration. In these cases one should proceed slowly, clear the vitreous from the anterior chamber and avoid any traction on the vitreous base or excessive hydration of the vitreous.

Lens dislocation may occur rarely into the vitreous cavity. It can be a total lens dislocation due to zonular weakness or just a subluxation because of undue pressure on the zonule. Even the nuclear fragments may get posteriorly dislocated in the event of posterior capsular tear.

Expulsive hemorrhage, though rare, can occur during or soon after the cataract surgery. Hypertension, arteriosclerosis, diabetes and raised intraocular pressure are known risk factors. Severe ocular pain, soakage of the eye pad and prolapse of the vitreous and the uveal tissue in the wound are the presenting features. Suprachoroidal drainage of the blood and reformation of the anterior chamber may salvage the eye. In hopeless cases evisceration of the eyeball becomes necessary.

Postoperative Complications

Depending on the time of occurrence, the postoperative complications of cataract surgery may be divided into two categories—early and late.

Early Postoperative Complications

Early postoperative complications of cataract surgery are listed in Table 27.3.

Table 27.3: Early postoperative complications of cataract surgery

1. Striate keratitis
2. Corneal edema
3. Prolapse of iris
4. Hyphema
5. Anterior uveitis
6. Delayed formation of the anterior chamber
7. Early endophthalmitis

Striate keratitis develops due to damage to the corneal endothelium during excessive manipulation within the anterior chamber or by prolonged and repeated irrigation. It usually disappears within a few days, but causes significant loss of corneal endothelial cells.

Corneal edema may be caused by surgical trauma, prolonged intraocular irrigation, pre-existing corneal endothelial dysfunction and raised IOP. The endothelial damage results in corneal edema.

The incidence of *prolapse of the iris* (Fig. 27.32) is grossly reduced owing to the use of sutures and construction of self-sealing incision. Once it is noticed without any signs of infection it must be reposed taking aseptic precautions and additional stitches are placed to repair the wound.

Postoperative hyphema usually appears on the fifth day due to leakage from the newly formed vessels in the section. Mydriatic-miotic instillations, topical corticosteroids and oral serratiopeptidase may be helpful in the absorption of blood. Massive hyphema needs paracentesis. When hyphema is

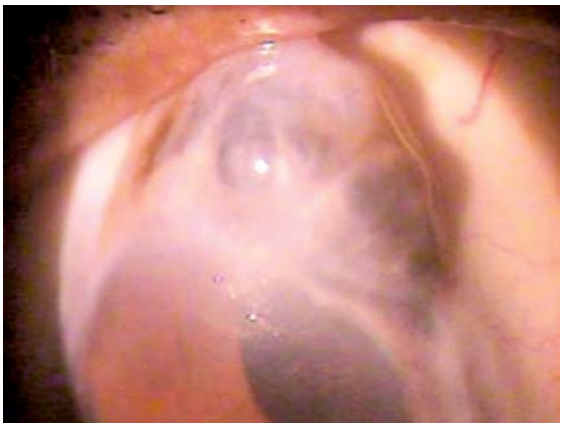


Fig. 27.32: Massive iris prolapse after ICCE

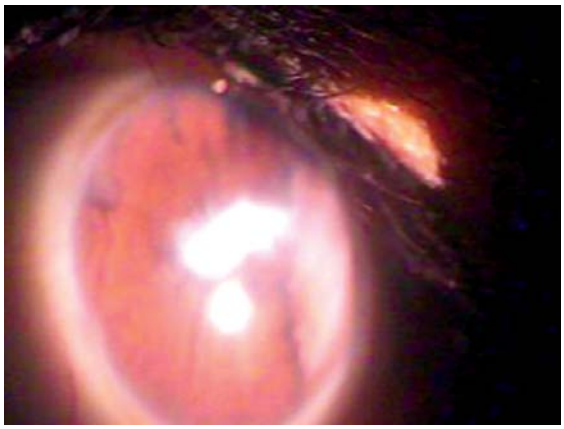


Fig. 27.33: Postoperative phthisis bulbi

associated with raised intraocular pressure, blood-staining of the cornea may develop. The pressure must be lowered by oral acetazolamide.

Mild anterior uveitis occurs in almost all cases of extracapsular lens extraction, therefore, postoperative topical mydriatic and corticosteroids should continue for some weeks. *Moderate to severe anterior uveitis* may occur in those patients who have diabetes and rheumatism. In such cases routine treatment of the disease should be supplemented with subconjunctival corticosteroid injections.

The *delayed formation of the anterior chamber* is caused by a ragged section, improper apposition of the wound, incarceration of the iris or the vitreous in the lips of the wound and detachment of the choroid. It can be prevented by following the correct surgical technique and proper suturing of the wound. The administration of acetazolamide or injection of air into the anterior chamber may restore the depth of the anterior chamber if the closure of the wound is proper.

Endophthalmitis after cataract surgery has been significantly reduced by the use of aseptic surgical techniques and antibiotics. Once the wound is infected the infection travels inside the eye and endophthalmitis supervenes. The wound infection often develops on the 2nd or 3rd day after surgery. It is marked by severe pain, edema of lids, chemosis of the conjunctiva, corneal haze, hypopyon, acute uveitis and dull fundus glow. Mild infection can be managed by intravitreal injection of antibiotics, systemic antibiotics and corticosteroids, or vitrectomy. In spite of aggressive treatment some eyes may end up in phthisis bulbi (Fig. 27.33). Severe infection often leads to panophthalmitis. The eye is often lost and has to be eviscerated.

Late Postoperative Complications

Secondary glaucoma, cystoid macular edema (CME), after cataract, retinal detachment and late onset endophthalmitis are late complications of cataract surgery (Table 27.4).

Table 27.4: Late postoperative complications of cataract surgery

1. Secondary glaucoma
2. Bullous keratopathy
3. Cystoid macular edema
4. Posterior capsule opacification or after cataract (in ECCE)
5. Retinal detachment
6. Late endophthalmitis

Secondary glaucoma may supervene due to the formation of peripheral anterior synechiae as a result of shallowing of the anterior chamber. Sometimes, the lens matter and particulate material from iridocyclitis block the trabecular meshwork and cause elevation of the intraocular pressure. The forward bulge of the vitreous can cause *pupillary block glaucoma*.

Bullous keratopathy is caused by a prolonged contact of the vitreous with the corneal endothelium, it may be associated with *cystoid macular edema (Irvine-Gass syndrome).*

Posterior capsule opacification (Fig. 27.34) or a thick after cataract is common after ECCE particularly in young patients (Fig. 27.35). It can be incised or dealt with Nd:YAG laser capsulotomy.

Retinal detachment occurs with greater frequency in aphakic eyes. Intraoperative vitreous loss, lattice degeneration, traumatic cataract and high myopia are risk factors. The detachment must be managed as early as possible.

Late onset postoperative endophthalmitis may occur due to infection by organisms of low virulence or fungal infection. Toxic reaction to intraocular lens material also presents as late endophthalmitis. *Propionibacterium acnes* infection manifests as a dense white plaque on the posterior capsule. Posterior capsulotomy, especially by Nd:YAG laser, would cause access of *P. acnes* into the vitreous cavity leading to endophthalmitis.

The relative merits and demerits of ECCE and ICCE are listed in Table 27.5.

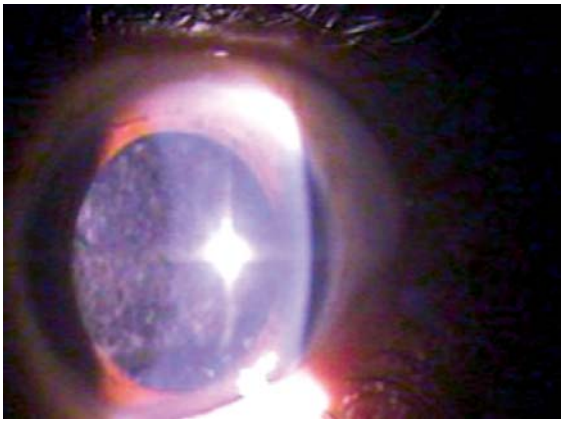


Fig. 27.34: Posterior capsule opacification

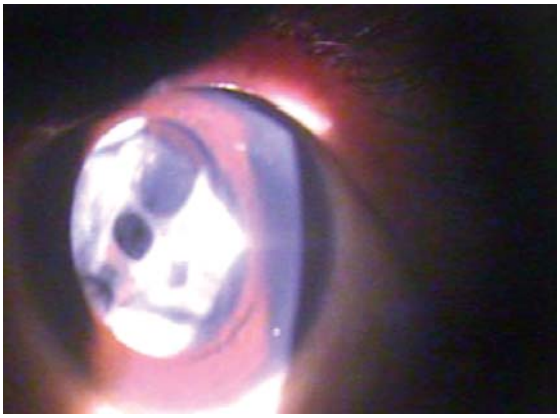


Fig. 27.35: Thick posterior capsule opacification

Intraocular Lens Implantation

Intraocular lens (IOL) implantation has become an established surgical procedure in the management of cataract. The main advantage of lens implantation is the restoration of vision, both central and peripheral, approximating to the precataractous level. The procedure eliminates all the major disadvantages of aphakia. The lens implantation may be performed soon after the delivery of the nucleus in cataract surgery, *primary implantation*, or any time postoperatively, *secondary implantation*.

Indications

1. Monocular cataract with good visual acuity in the other eye
2. Cataract patients with intolerance for contact lens
3. Patients with macular degeneration or retinitis pigmentosa
4. High degree of aniseikonia
5. Mentally handicapped patients.

Table 27.5: Merits and demerits of ECCE and ICCE

	ECCE	*ICCE*
1. Technique	Relatively difficult to perform and time consuming	Easy to perform and less time consuming
	Requires operating microscope	Does not require operating microscope
2. Limitations		
a. Age	Can be performed at all ages	Cannot be performed below the age of 20 years as the zonule is tough
b. Luxated lens	Cannot be removed by this procedure	Can be removed by this procedure
3. PC IOL implantation	'In-the-bag' implantation is possible	'In-the-bag' implantation is not possible
4. Intraoperative procedure		
a. Limbal incision	Small for the delivery of the nucleus	Large incision for the delivery of the lens in toto
b. Capsulotomy	Needed	Not needed
c. Sutures	Usually not required	Usually required
5. Intraoperative vitreous loss	Rare as the posterior capsule is intact	May occur as there is no barrier (capsule)
6. Postoperative complications		
a. Postoperative capsular opacification (PCO)	Often occurs and needs capsulotomy	No chance of PCO, second operation not needed
b. Anterior uveitis	May occur	Rarely occurs
c. Retinal detachment	Incidence is insignificant	Relatively higher incidence
d. Cystoid macular edema	Less common	More frequent
e. Astigmatism	Low due to small incision	High due to bigger incision
7. Wound	Small, stable and heals early	Large wound that takes time to heal
8. Visual rehabilitation	Occurs early	Takes longer time

In spite of the fact that IOL implantation is a safe and popular technique, there are special problems associated with it. It should, therefore, be not performed in some categories of patients.

Contraindications

1. Uncontrolled and recurrent ocular inflammations
2. Corneal endothelial decompensation or low endothelial cell count
3. Nonmotivated patients.

Types of IOL Implantation

The IOL implantation is mainly of two types:

1. Anterior chamber IOL implantation (ACIOL)
2. Posterior chamber IOL implantation (PCIOL).

Anterior Chamber Lenses

The anterior chamber lenses (ACL) lie in front of the pupil and the iris and are either supported by the scleral spur in the angle of the anterior chamber or by the iris. Out of several types and styles, the Kelman style with four point fixation (Fig. 27.36) is preferred. The ACLs are not free from complications and their long-term results may not be satisfying. Therefore,they are practically discarded for primary implantation. However, there remains a number of situations when ACL implantation is indicated.

Indications of ACL Implantation

1. After ICCE
2. Ocular trauma with inadequate posterior capsular support

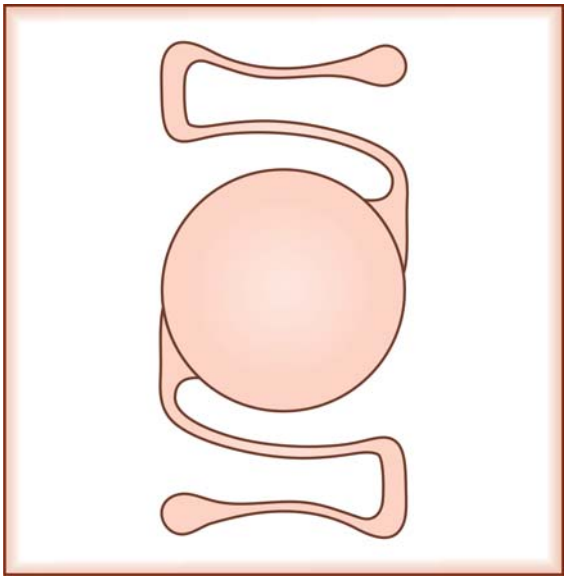


Fig. 27.36: Anterior chamber lens

3. After ECCE with vitreous loss and insufficient capsular support
4. Phakic ACL to correct high myopia.

Technique of ACL Implantation

Before commencing the implantation either after ICCE or ECCE good miosis is achieved and the anterior chamber is filled with viscoelastic material. The ACL is held by the optic with McPherson forceps and inserted so that the distal feet lie on the scleral spur (Fig. 27.37). To insert the proximal feet, the haptic is pushed backwards after slightly retracting the wound.

For implanting the Worst iris claw lens (Fig. 27.38), the lens is held over the surface of the iris and centered. The iris fold below the claw is then grasped with enclavation forceps and the lens is depressed onto the forceps. The same procedure is repeated on the other side.

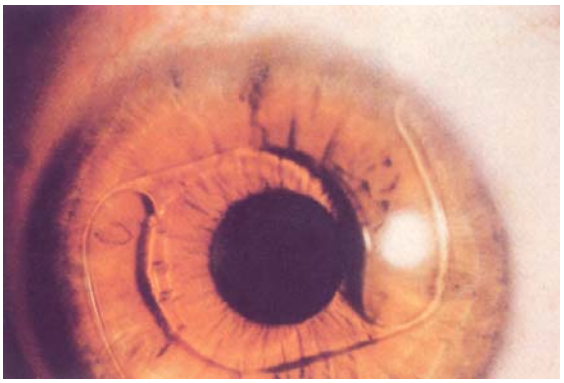


Fig. 27.37: Kelman anterior chamber lens

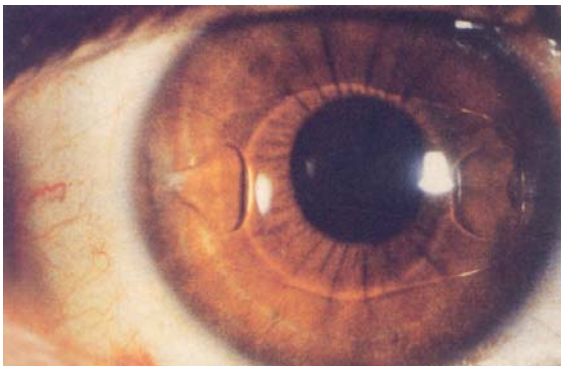


Fig. 27.38: Worst claw lens

Complications

The ACL implantation may lead to a number of complications (Table 27.6). They include corneal endothelial damage, corneal decompensation or bullous keratopathy (Fig. 27.39), pupillary tuck, uveitis, glaucoma and hyphema (UGH) syndrome and cystoid macular edema. Sometimes the implanted lens becomes unstable and gets subluxated.

Posterior Chamber Lenses

Posterior chamber lenses (PCL), both single piece or multipiece with J-shaped or C-shaped loops, are available (Fig. 27.40). Currently, biconvex optics is preferred over planoconvex.

For small incision cataract surgery with or without phacoemulsification, foldable silicone, acrylic or hydrogel lenses are used (Fig. 27.41). To

Table 27.6: Complications of ACIOL implantation

1. Corneal endothelial damage
2. UGH syndrome
3. CME
4. Pupillary tuck
5. Decentration of the lens

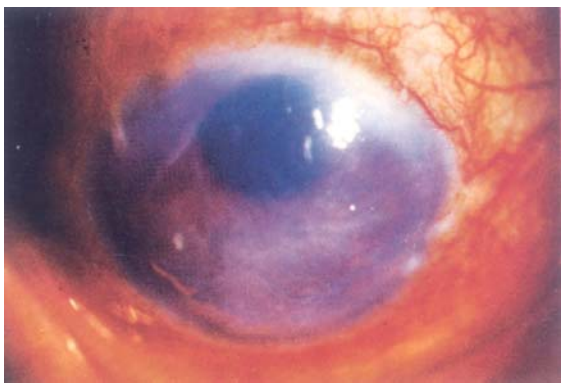


Fig. 27.39: Bullous keratopathy

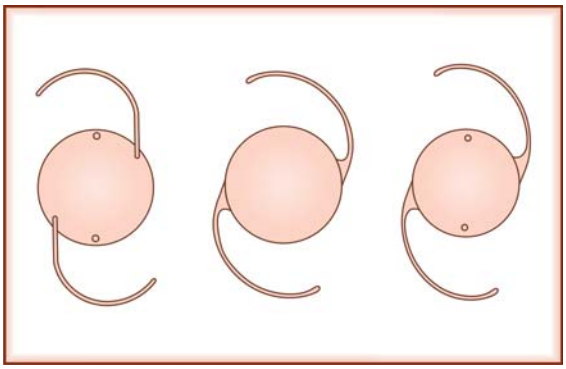


Fig. 27.40: Posterior chamber IOLs

Fig. 27.41: Foldable IOL (*Courtesy*: Alcon)

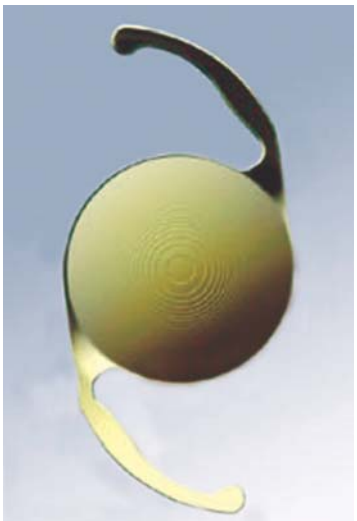


Fig. 27.42: Multifocal IOL (*Courtesy*: Alcon)

obtain accommodation, multifocal and accommodative IOLs have been developed (Fig. 27.42).

The posterior chamber IOL can be implanted either in the ciliary sulcus or in the capsular bag. The 'in-the-bag' implantation is more physiological and usually preferred.

Technique of PCL Implantation

After meticulous aspiration of the cortex in ECCE, the anterior chamber and capsular bag are filled with viscoelastic material. The PCL is grasped by the optic with a lens holding forceps, and the inferior haptic and optic slid into the capsular bag (video). The superior haptic is grasped with McPherson forceps, flexed behind the optic and gently placed under the upper flap of the capsule. The lens is dialed to the horizontal position and wound is closed.

Complications

The PCL implantation offers better visual results with a relatively lesser rate of complications (Table 27.7).

Table 27.7: Complications of PCIOL implantation

1. Pupillary capture
2. Decentration
3. Posterior dislocation of IOL
4. Posterior capsular rent
5. Posterior capsule opacification
6. CME

Pupillary capture occurs when the lens optic lies anterior to the iris (Fig. 27.43). The lens may get decentered. When the IOL is decentered inferiorly it is called *sunset syndrome* (Fig. 27.44) and superiorly, *sunrise syndrome*. Rarely the lens is dislocated posteriorly and floats in the vitreous.

In case of inadvertent *posterior capsular rent* with the loss of adequate support, a scleral fixated PCL can be placed. After a meticulous anterior vitrectomy the lens is implanted in the ciliary sulcus and sutured to the sclera by 9-0 or 10-0 polypropylene suture.

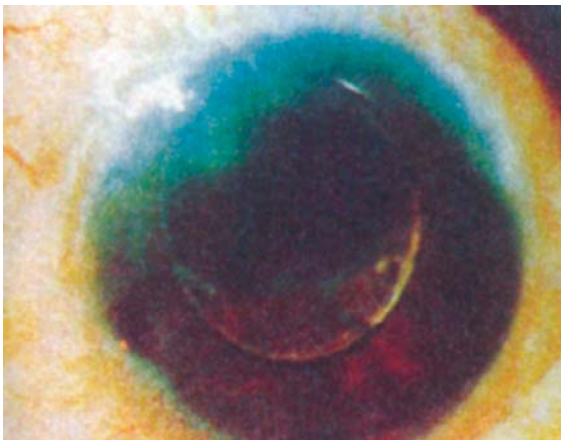


Fig. 27.43: Pupillary capture (*Courtesy*: Dr Abhay R Vasavada, Ahmedabad)

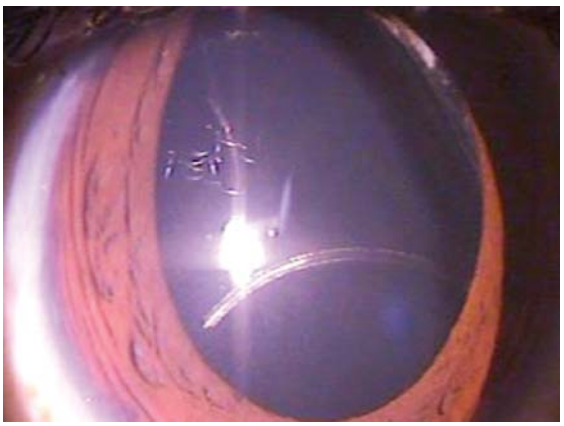


Fig. 27.44: Sunset syndrome

The most common delayed complication of ECCE is *posterior capsule opacification* (PCO) which results from the proliferation and migration of remnant lens epithelial cells. Nd:YAG laser posterior capsulotomy restores the vision in such cases. CME may develop in some of the cases of PCL implantation.

PARS PLANA SURGERY

The pars plana surgery is a technique by which the lens or the vitreous is removed through a small incision in the region of pars plana. A sophisticated automated suction-cutting device is used for this purpose while maintaining the normal intraocular pressure. The technique avoids damage to the corneal endothelium and causes least damage at the site of entry into the eye. Pars plana surgery can be performed either for the anterior segment disorders or for the posterior segment diseases.

Indications

The indications for pars plana surgery are:

1. Occlusio pupillae
2. Congenital or traumatic cataract
3. Traumatic vitreous incarceration
4. Complicated retinal detachment
5. Non-clearing vitreous hemorrhage
6. Posterior segment intraocular foreign body (video)
7. Endophthalmitis
8. Intravitreal or subretinal cysticercus cyst (video).

Patients with complicated retinal detachment, especially with giant retinal tears and marked proliferative vitreoretinopathy, require trans pars plana vitrectomy (TPPV). Advanced vitrectomy removes vitreous opacities, relieves vitreoretinal traction and reattaches the retina by internal tamponade. Air, inert expansile gases such as sulphur hexafluoride (SF6) or perfluoropropane (C3F8), and silicone oil are used as tamponading agents for internal closure of the retinal breaks.

The TPPV helps in clearing the organized vitreous hemorrhage, removing the fibrovascular membrane either by delamination or by segmentation, and relieving the traction on retina. It can be followed by endophotocoagulation.

Pars plana vitrectomy basically has two main purposes:

1. Therapeutic, and
2. Diagnostic.

Therapeutic vitrectomy is done for debulking the vitreous of debris, pus, blood and pathogenic organisms.

Diagnostic vitrectomy is mainly performed for obtaining the sample for culture and sensitivity tests in patients with endophthalmitis.

RETINAL REATTACHMENT SURGERY

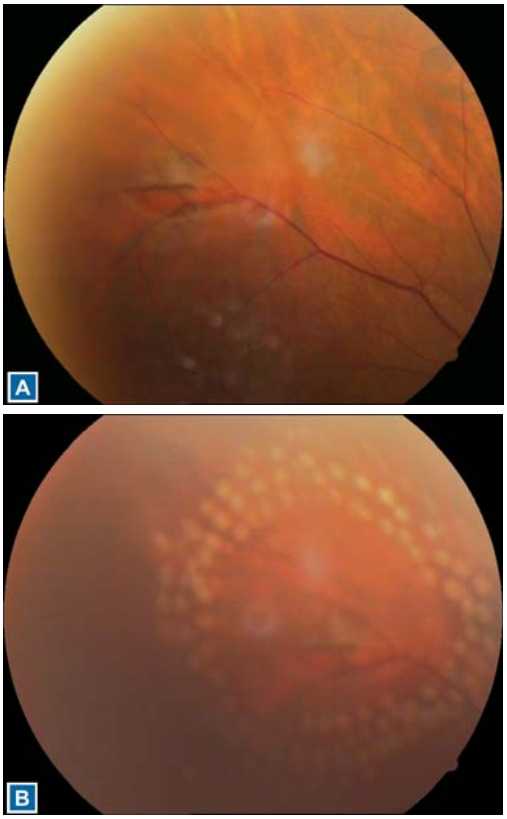
The retinal reattachment surgery can be performed under local or general anesthesia. Topical anesthetic drops are not used as they cloud the cornea and interfere with the ophthalmoscopic examination.

The basic principle of a successful RD surgery is to see, to seal and to support the retinal break. An accurate localization of retinal breaks is carried out by an indirect ophthalmoscopic examination.

After a 360° peritomy, bridle sutures are passed through the four rectus muscles (video). One must localize all the breaks and the sclera underlying them is marked by diathermy. A firm adhesion between the sensory retina and the RPE or Bruch's membrane is required to seal the break (Figs 27.45A and B). This can be achieved by cryopexy (–70°C) or photocoagulation. Indenting the sclera and choroid towards the retinal break (scleral buckling) supports the break and facilitates settling of the detached retina. It also relieves the vitreous traction.

Silicone tyres and sponges are used as an explant (episcleral placement) or an implant (intrascleral placement) for scleral buckling. 4-0 or 5-0 polybutylate-coated braided polyster (Ethibond) or siliconized silk suture material is used to hold the buckle in place. Depending on the orientation a buckle can be either circumferential or radial. Some surgeons prefer to do an encircling procedure routinely wherein a silicone strap provides a 360° indentation (Fig. 27.46).

The indications of subretinal fluid (SRF) drainage in the RD surgery include long-standing detachment, bullous detachment, and eyes wherein there is a danger of rise in intraocular pressure. The post drainage hypotonia is countered by intravitreal injection of Ringer's lactate or air.



Figs 27.45A and B: (A) Retinal break, (B) Sealed retinal break by photocoagulation (*Courtesy*: Prof YR Sharma, Dr RP Center, New Delhi)

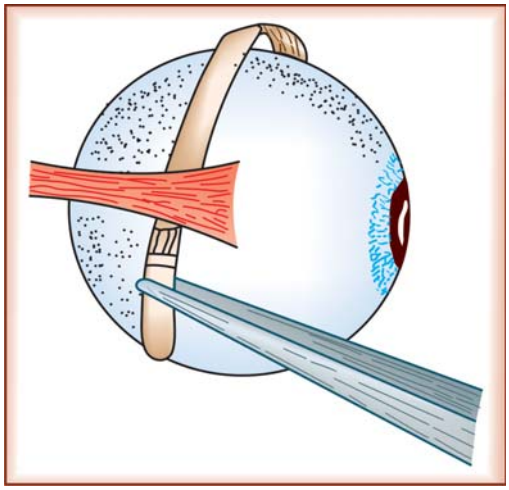


Fig. 27.46: Encircling procedure

Prognosis

The surgical results of rhegmatogenous detachment of retina are good. If attended early and operated properly, anatomic reattachment is achieved in at least 90% of cases. Late and untreated cases have a poor prognosis.

Complications

Choroidal or vitreous hemorrhage, choroidal detachment, elevation of intraocular pressure and nonattachment of the retina are some of the complications of RD surgery.

Prophylactic photocoagulation or cryopexy is recommended in fellow eyes with retinal breaks of the patient with RD, and myopes with atrophic holes and tears.

EVISCERATION OF THE EYEBALL

Indications

Evisceration is recommended in the following conditions:

1. Panophthalmitis
2. Anterior staphyloma
3. Expulsive hemorrhage.

Procedure

Evisceration can be done either under general or local anesthesia. The lids are retracted by an eye speculum and a stab incision is made at the limbus (video). The cornea is removed with the scissors and all the intraocular contents (uvea, lens, vitreous and retina) are scooped out with an evisceration spoon. The inner surface of the sclera is cleaned with a swab and the cavity is sprayed with appropriate antibiotic.

For obtaining satisfactory cosmetic results a silicone ball of about 16 mm diameter is inserted within the scleral cup provided there is no active inflammation. The sclera, Tenon's capsule and the conjunctiva are stitched. After 3 weeks of operation, a cosmetic shell can be fitted.

ENUCLEATION OF THE EYEBALL

Indications

Enucleation of the eyeball is indicated in the following conditions:

1. Painful blind eye
2. Intraocular tumors (retinoblastoma, malignant melanoma)
3. Perforating injury to the eye with loss of vision
4. Large retained intraocular foreign body with teethered globe
5. Anterior and ciliary staphyloma in a blind eye.

Procedure

General anesthesia is preferred for enucleation. An eye speculum is inserted (video). The conjunctiva is incised around the limbus. It is undermined up to the insertion of the extraocular muscles. The insertions of all the four rectus muscles are defined and a silk suture is passed near the insertion of each muscle. Then a muscle hook is passed under the tendon and one-by-one the rectus muscles are severed. The speculum is pressed backward to obtain a forward bulge of the eyeball. The eyeball is rotated medially and the enucleation scissors is introduced behind the eye until the optic nerve is felt. The nerve is engaged between the blades of the scissors and severed. Improper positioning of the blades of scissors may cause perforation of the globe. The eyeball is drawn forward and the attached tendons of the superior and inferior oblique muscles are severed.

When enucleation is performed for malignancy of the eye, a bigger stump of the optic nerve should be obtained for histopathological examination to assess the extension of the growth. The cutting of the optic nerve is usually accompanied by bleeding which is controlled by packing the cavity with gauze wrung in hot saline. The edges of the conjunctiva are sutured. After application of an antibiotic ointment the lids are closed.

For giving near natural mobility to an artificial eye, an acrylic implant is inserted soon after the removal of the eyeball and the rectus muscles are sutured over it followed by conjunctival sutures. Extrusion of the implant is not rare.

EXENTERATION OF THE ORBIT

The *en masse* removal of the contents of the orbit along with the eyeball is known as *exenteration of the orbit*. It is done for the malignant tumors of the orbit and in the extraocular stage of intraocular neoplasms. The operation is performed under general anesthesia. The exenterated orbit may be covered by a prosthesis attached to the spectacle frame.

STRABISMUS SURGERY

The purpose of strabismus surgery is not only to align the two eyes cosmetically but to restore the binocularity as well. As a rule, the surgery should be performed as soon as the child is old enough to cooperate in orthoptic exercises. Postponement of operation may lead to the development of amblyopia and failure to restore binocular vision. In long-standing strabismus, surgery is done mainly for cosmetic purpose.

For obtaining the alignment, the underacting muscle is strengthened by resection and overacting muscle is weakened by recession. Free tenotomy or guarded tenotomy can also weaken the overacting muscle. In resection the length of the tendon of the muscle is shortened to enhance its action. In recession, the muscle is detached from its insertion and reattached a few millimeters behind its insertion to weaken its action.

Convergent strabismus is usually undercorrected, while divergent strabismus is fully corrected or over-corrected. The medial rectus recession gives more correction than the resection, and the lateral rectus resection often results in more correction of strabismus than the recession.

The medial rectus should not be recessed more than 5 mm otherwise the patient may develop discomfort during reading and near work owing to convergence weakness. When the deviation is more than 10°, surgery on both medial and lateral rectus muscles may become necessary. To correct large degrees of deviation, besides operating the two muscles of one eye, the muscles of other eye are also tackled.

Recession of Medial Rectus

General anesthesia is preferred in a child. An eye speculum is inserted, a fixation suture is passed at the medial limbus and the eye is rotated laterally. The conjunctiva is incised vertically just medial to the traction suture and the incision is extended concentrically with the limbus upwards and downwards. The conjunctiva is undermined towards the inner canthus. Tenon's capsule is cut along the upper and the lower border of the muscle and a hook is passed under the tendon of the medial rectus. The muscle is freed from its lateral expansions as well as from the episclera. The calipers are set to the desired length of recession, and the distance is measured from the insertion of the tendon and marked on the sclera near the upper and lower borders of the muscle with a diathermy. Two 6-0 polyglycolic acid or catgut sutures are passed through the upper and lower edges of the tendon 2 mm behind the insertion. The tendon is divided at its insertion by cutting with a scissors, and the sutures are passed through the superficial layers of the sclera at right angles to the long axis of the muscle at the points already marked (Fig. 27.47). The sutures are tied and conjunctival incision is closed.

Resection of Lateral Rectus

Resection of the lateral rectus muscle is performed by rotating the eye medially by a suture placed at the lateral limbus. A conjunctival incision is made 2 mm lateral and concentric with the limbus and the conjunctiva is undermined temporally. Slit

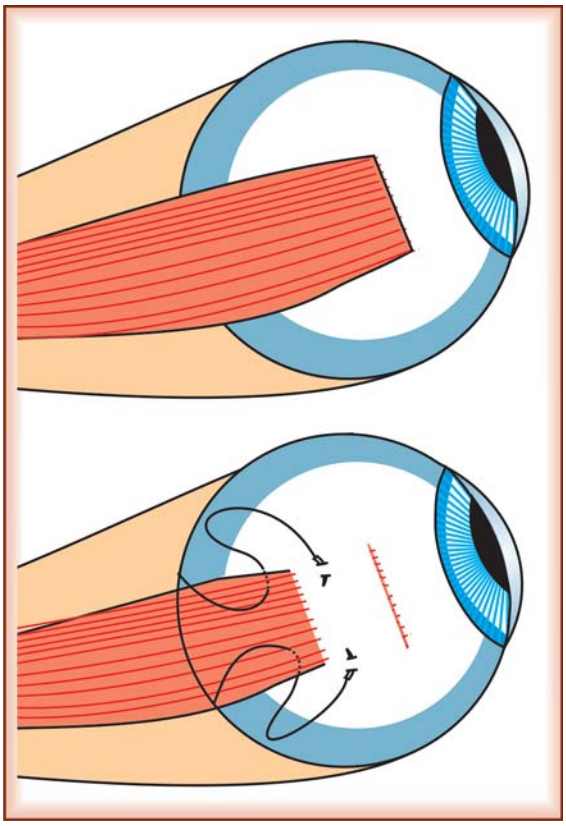


Fig. 27.47: Recession of the medial rectus

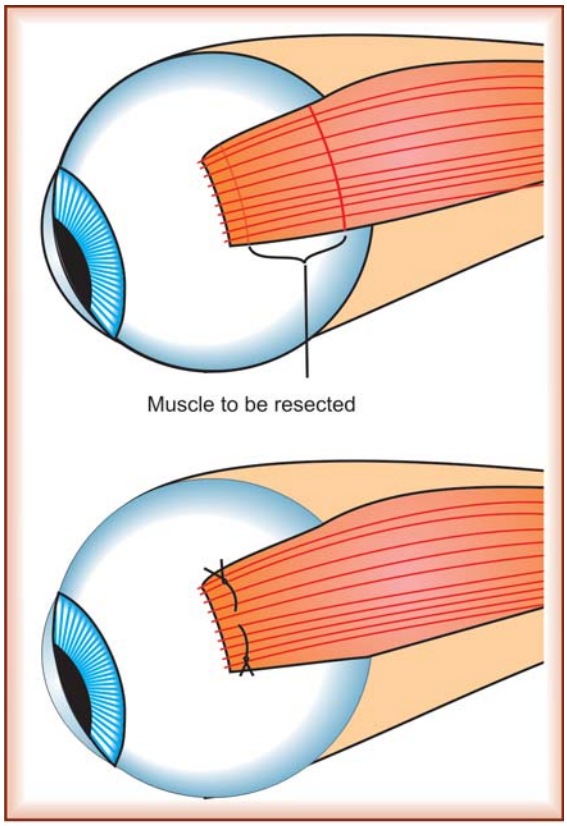


Fig. 27.48: Reattachment of the lateral rectus after resection

incisions are made in Tenon's capsule and the lateral rectus is exposed. The desired length of the muscle to be resected is measured with the help of calipers and marked on the muscle itself. Two whip sutures are passed through the upper and lower edges of the muscle nearly 1 mm behind the mark. The muscle is divided at the marked site as well as at its insertion. The sutures are then passed through half the thickness of sclera at the site of original insertion of the muscle and tied (Fig. 27.48). The conjunctival incision is closed.

Resection of medial rectus and recession of lateral rectus muscles (video) are performed to correct divergent strabismus.

Complications

Undercorrection, overcorrection, diplopia, enophthalmos, conjunctival cyst and granuloma are some of the complications of strabismus surgery. Encouraging results of strabismus surgery are obtained in accommodative esotropia and in prominent eyes.

SURGERY OF THE LACRIMAL PASSAGE

Syringing

Persistent epiphora in a newborn occurs due to obstruction of the lower end of the nasolacrimal

duct. The obstruction may be confirmed as well as overcome in some cases by syringing and probing.

Syringing is performed under general anesthesia in a child. The lower punctum is dilated with a punctum dilator. A lacrimal canula attached to a syringe is inserted through the punctum and the canaliculus, and the passage is syringed with slight pressure.

Probing

When the syringing fails to overcome the obstruction, probing is planned.

The canula used for syringing is removed and a probe of small size is passed through the dilated punctum along the canaliculus into the sac where some resistance is felt. Then the probe is swung into vertical position and passed downwards, backwards and laterally through the nasolacrimal duct into the inferior meatus of the nose (Fig. 27.49). Subsequent syringing will verify the patency of the lacrimal passage. Turbinate fracture, as a part of probing, may be performed using a large size Bowman's probe.

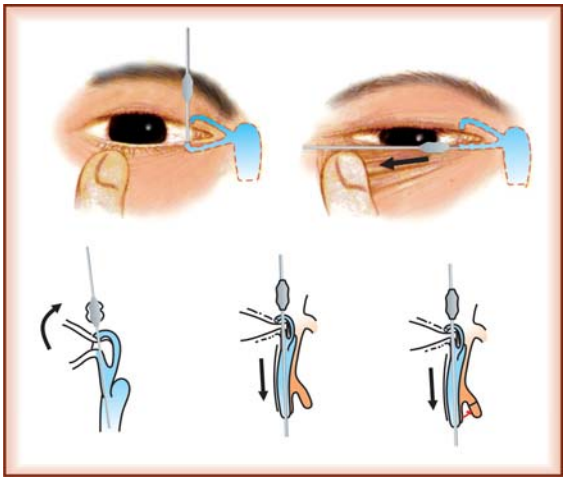


Fig. 27.49: Procedure of probing
(*Courtesy*: Drs. Vikas Mahatme and Chitra Pande, Nagpur)

Balloon Dacryoplasty

Balloon catheter dilatation is an effective procedure in removing the obstruction of the nasolacrimal duct especially in congenital dacryocystitis.

Lacrimal Intubation

When repeat probing fails, silicone tube intubation is indicated.

Dacryocystectomy

The removal of the sac is known as *dacryocystectomy*.

Indications

The dacryocystectomy is indicated in following conditions:

1. Tuberculosis of the sac
2. Malignancy of the sac
3. Chronic fibrotic dacryocystitis with or without atrophic rhinitis.

Procedure

The operation is done under general anesthesia in children and under local infiltration anesthesia in adults. Xylocaine 2% with adrenaline is injected at the junction of inferior orbital margin and anterior lacrimal crest. The solution is also infiltrated in the region of medial palpebral ligament, the nasolacrimal duct and posterior lacrimal crest.

About 2 cm long skin incision is made 3 mm medial to the inner canthus (Fig. 27.50A). The orbicularis oculi fibers are split (Fig. 27.50B) and the margins of the wound are retracted by cat's paw retractor. The anterior lacrimal crest is defined with the help of a blunt dissection and the attachment of the medial palpebral ligament to the anterior lacrimal crest is divided to expose the

lacrimal sac. Thereafter, the attachments of the sac with the lacrimal fossa are freed. The sac is drawn forward and twisted and is severed from the nasolacrimal duct. The upper end of the duct is curetted and the incision is closed by sutures.

Excessive bleeding may be encountered during the operation owing to damage to the angular vein. Postoperative epiphora is not unusual.

Dacryocystorhinostomy

In dacryocystorhinostomy (DCR) a communication is established between the sac and the middle meatus of the nose to bypass the obstructed nasolacrimal duct.

Indications

DCR is indicated in the following conditions:

1. Chronic dacryocystitis
2. Mucocele of the lacrimal sac.

Contraindications

In the presence of a gross nasal pathology, atrophic rhinitis, nasal polyp and lupus, the operation is contraindicated. Marked fibrotic sac, osteomyelitis of the lacrimal fossa and tuberculous dacryocystitis are other conditions where the operation is deferred.

Procedure

Usually, the operation is performed under local anesthesia. The nasal cavity is packed with a gauze soaked in 4% solution of xylocaine with adrenaline (video). The skin incision and exposure of the sac are same as in dacryocystectomy.

The periostem is separated (Fig. 27.50C), and the lacrimal fossa is exposed (Fig. 27.50D). The overlying periosteum is lifted and a small opening (12 × 10 mm) is made with a bone punch to expose the nasal mucosa (Fig. 27.50E). A vertical incision is made in the medial wall of the lacrimal sac to form the anterior and posterior flaps. Then an H-shaped incision is made in the nasal mucosa with the horizontal incision in the middle to raise 2 flaps of the mucosa (Fig. 27.50F).

The posterior flap of the sac is sutured to the posterior flap of the nasal mucosa (Fig. 27.50G) and the anterior sac flap to the anterior nasal flap using 6-0 polyglycolic acid suture (Fig. 27.50H). The divided ends of the medial palpebral ligament are reposited and sutured, the split margins of orbicularis oculi muscle sutured (Fig. 27.50I), and the skin incision is closed. The wound is dressed with antibiotic ointment and bandaged.

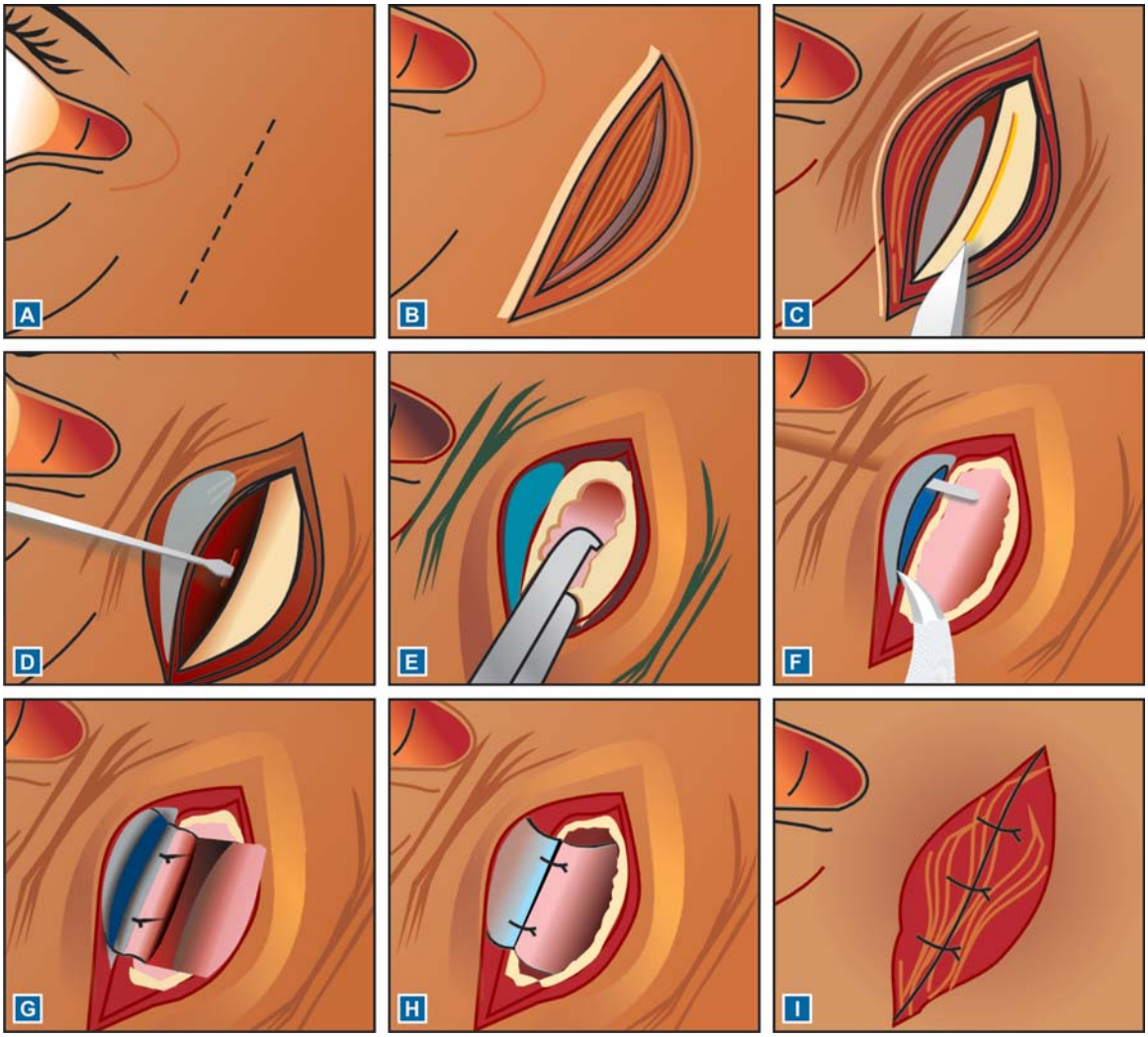
The nasal pack is removed on the second day of operation. The syringing is done on the third postoperative day and the skin sutures are removed on the seventh day.

Complications

Complications of dacryocystorhinostomy are not frequent. Bleeding may occur from an injury to the angular vein or vascular nasal mucous membrane. The formation of clot may obstruct the communication. Postoperative infection is rarely seen.

Other Procedures

Endonasal DCR (video), laser-assisted DCR, and transcanalicular laser DCR are some of the alternatives to the conventional external DCR. When both upper and lower canaliculi are blocked, conjunctival DCR is performed. In this procedure a free communication is established between the lacrimal lake and the middle meatus of the nose through Jones pyrex glass tube.



Figs 27.50A to I: Steps of dacryocystorhinostomy. (A) Site of skin incision, (B) Separation of orbicularis muscle fibers, (C) Periosteal separation, (D) Exposure of lacrimal fossa, (E) Making bony osteum, (F) Flaps of sac raised, (G) Suturing of posterior flaps, (H) Suturing of anterior flaps, (I) Suturing of orbicularis (*Courtesy*: Drs. Vikas Mahatme and Chitra Pande, Nagpur)

CRYOTHERAPY OR CRYOPEXY

Cryopexy involves application of freezing cold by a cryoprobe attached to a cryomachine (Figs 27.51 and 27.52). Liquid nitrogen or nitrous oxide or carbon dioxide is used as a coolant. The size of the tip of the probe varies. One mm diameter tip is used for vitreous surgery, 1.5 mm tip size for cataract extraction, 2.5 mm for retina and choroid and 4 mm for cyclocryopexy.

Cryopexy may be performed under topical or infiltrative or general anesthesia.

Mode of Action

Rapid freezing produces both extracellular and intracellular ice crystals disturbing the cell

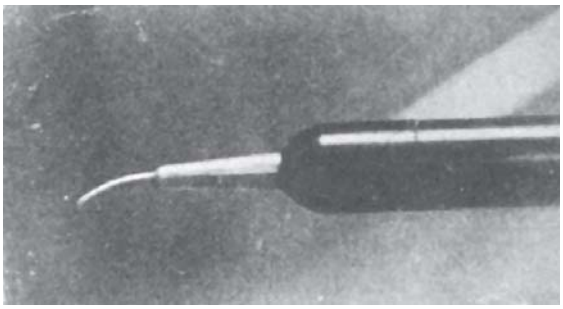


Fig. 27.51: Ophthalmic cryoprobe

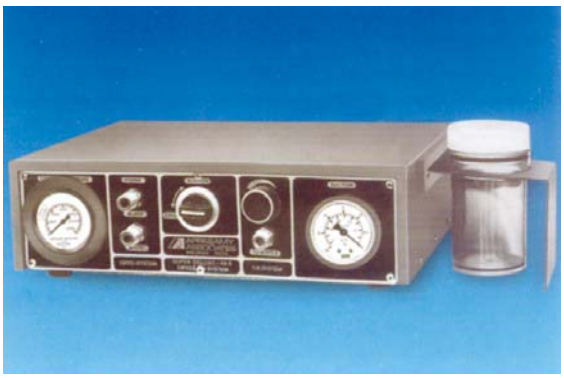


Fig. 27.52: Ophthalmic cryo unit

membranes and organelles. Repeated freezing-thawing can produce tissue adhesion, vascular occlusion and tissue necrosis.

Indications

Common indications of cryo therapy are given below.

1. *Intracapsular cataract extraction*: A –35 or –40° C temperature induces a firm adhesion between the crystalline lens and the cryoprobe for easy delivery of the lens.
2. *Retinal detachment:* Cryopexy seals the break in the retina. It is also used for prophylactic purpose for the treatment of areas of retinal degenerations and breaks.
3. *Neovascular and absolute glaucomas:* Cryo application to the ciliary body reduces the intraocular pressure by destruction of the ciliary epithelium.
4. *Tumors*: Cryotherapy may be used for the treatment of small solitary hemangioma, basal cell carcinoma and retinoblastoma.
5. *Retinopathy of prematurity*: Cryotherapy is often used in the management of retinopathy of prematurity.
6. *Miscellaneous*: Cryopexy may be used for the management of giant papillae of vernal kerato-conjunctivitis, molluscum contagiosum, pars planitis and advanced cases of proliferative diabetic retinopathy.

LASER THERAPY

LASER is an acronym for Light Amplification by Stimulated Emission of Radiation. The useful properties of a laser beam are monochromatism, coherency, collimation and concentration in a short time. The lasers used for ophthalmic purposes are listed in Table 27.8.

The laser can be delivered to the eye by a slit-lamp, an indirect ophthalmoscope or by a fiberoptic probe (endophotocoagulation) during trans pars plana vitrectomy.

Table 27.8: Lasers for ophthalmic use

Name	*Type*	*Wavelength*	*Use*
Argon	Gas	488-515 nm	Photocoagulation
Krypton	Gas	647 nm	Photocoagulation
Ruby	Solid	694 nm	Photocoagulation
Diode	Semi-conductor	780-850 nm	Photocoagulation
Nd:YAG	Liquid/solid	1062 nm	Photodisruption
Dye	Liquid	Multitude	Photoradiation
Excimer	Gas	193 nm	Photoablation
Carbon dioxide	Gas	10600 nm	Photovaporization

Photocoagulation

Photocoagulation is a thermal effect of laser and utilized for coagulation of new blood vessels (Fig. 27.53), formation of chorioretinal adhesion and destruction of the retinal pigment epithelium in certain conditions.

Indications

Common indications of photocoagulation are as follows:

1. Diabetic retinopathy
2. Branch or central retinal vein occlusion
3. Eales' disease
4. Retinal reattachment surgery
5. Retinopathy of prematurity
6. Tumors and cysts
7. Subretinal neovascular membrane (AMD, POHS)
8. Glaucoma: Laser is extensively used in reducing the IOP in glaucoma by (i) trabeculoplasty in POAG, (ii) iridotomy in PACG, especially to control an acute attack, and (iii) cyclophotocoagulation in neovascular and absolute glaucoma.
9. Miscellaneous: Laser can also be used for dilatation of pupil (photomydriasis), coreoplasty for updrawn pupil and suturolysis after trabeculectomy.

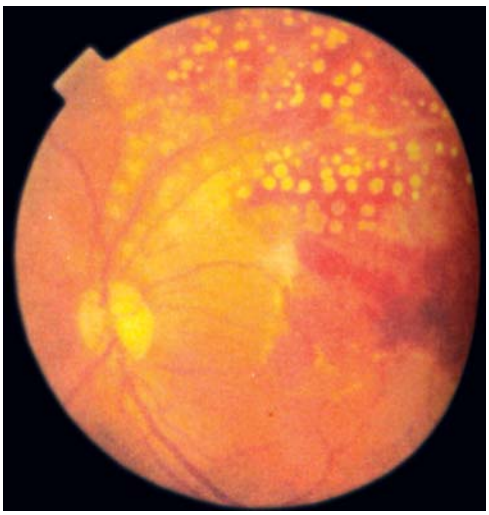


Fig. 27.53: Sector photocoagulation in BRVO

Photodisruption

Neodymium: Yttrium-Aluminum-Garnet (Nd: YAG) laser (Fig. 27.54), on focusing, forces away electrons from molecules producing plasma and shock waves which disrupt the ocular tissues. It is often employed in cutting the membrane or tissues in the eye.

Indications

1. Posterior capsule opacification or after cataract
2. Intravitreal membrane
3. Phacolysis (laser cataract surgery).

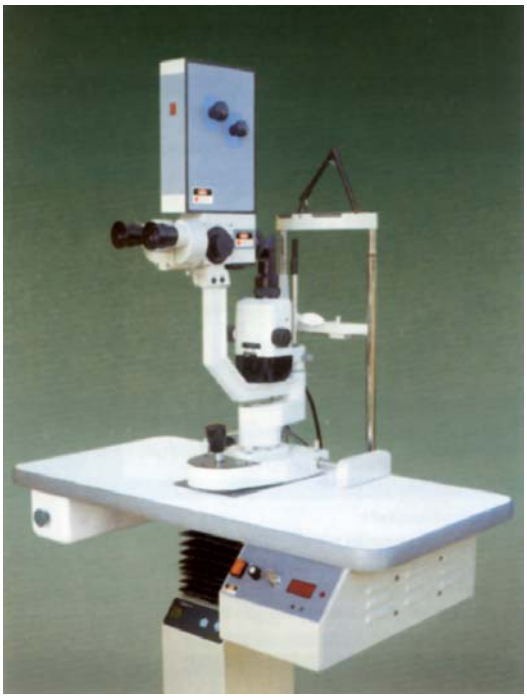


Fig. 27.54: Nd: YAG Laser
(*Courtesy*: Appasamy Associates)

Photoablation

The excimer laser is used for corneal modeling in refractive surgery (photorefractive keratectomy and LASIK). The excimer laser (Argon fluoride 193 nm) can break bonds of cells and reduce them to molecules that diffuse away in a short time.

Indications

1. *Photorefractive keratectomy* (PRK) is a technique to correct moderate degree of myopia (2-6 D). The corneal epithelium is initially removed. The excimer laser ablates the optical part of anterior stroma of the cornea. On healing the cornea gets flattened. The postoperative recovery is slow and the residual corneal haze may impair the vision in some cases.
2. *Laser in-situ keratomileusis* (LASIK) is a more refined procedure to correct myopia upto 16 D.

Procedure

The technique consists of three steps (video):

1. Creation of a flap of epithelium and superficial stroma
2. Ablation of mid-corneal stromal tissue (400 micron) by eximer laser, and
3. Reposition of the flap.

LASIK flattens the cornea without causing pain. Glare, corneal haze and under correction or over correction of myopia are some of the complications of the surgery.

Photodynamic Therapy

Photodynamic therapy (PDT) consists of two essential components, the photosensitizer (dye) that accumulates in the target tissue and a specific laser light corresponding to the absorption peak of the dye. Benzoporphyrin derivative, verteporfin, and a diode laser (690 nm) are used in photodynamic therapy to treat the subretinal neovascular membrane in diseases like age-related macular degeneration (AMD) and presumed ocular histoplasmosis syndrome (POHS).

The photosensitizer molecule is excited following the light absorption from the laser. The energy from the excited molecule is transferred to release free-radicals and production of singlet oxygen. Both the mechanisms destroy the subretinal neovascular-complex.

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CHAPTER 28 Ocular Manifestations of Diseases of the Central Nervous System

Eye is essentially a part of the central nervous system (CNS), therefore, its involvement in the diseases of the central nervous system is not uncommon. The presenting ocular signs may be of great diagnostic significance.

INFLAMMATORY DISORDERS

Meningitis

Acute suppurative meningitis frequently causes papillitis due to descending infection. There is always conjugate lateral deviation of the eyes. The paralysis of abducent nerve is common, third and fourth cranial nerves may also be involved. Moderate degree of bilateral papillitis or, occasionally, papilledema is found in tuberculous meningitis. The terminal cases of tuberculous meningitis may show small multiple choroidal tubercles. Chronic chiasmal arachnoiditis may cause bilateral primary optic atrophy.

Encephalitis

Diplopia and general lethargy are common symptoms of encephalitis. Ptosis is often present due to the involvement of third cranial nerve. Spasmodic conjugate upward movement of the eyeballs (*oculogyric crisis*) accompanied by synergic movements of head and neck occur in late stages of encephalitis. Paresis of accommodation and convergence may also be seen. Papillitis and papilledema are infrequent.

Brain Abscess

Cerebral abscess occurs more frequently than the cerebellar. Middle ear infection is the chief cause of cerebral abscess affecting the temporal lobe. The cerebellar abscess occurs even more frequently with otitis media. Nearly half of the cases of cerebellar abscess develop papilledema on the side of the abscess, and in bilateral papilledema the swelling is greater on the side of the abscess. This sign has a localizing value and differentiates the abscess from a tumor. Further, papilledema persists longer after the operation for an abscess than for a tumor or may even commence only after the surgery. Unilateral ptosis and mydriasis are pathognomonic of an ipsilateral cerebral or cerebellar abscess. Unilateral third nerve paralysis is quite frequent but is often incomplete. Partial third nerve paralysis with contralateral hemiplegia indicates abscess of the temporal lobe implicating the third nerve and the internal capsule by pressure. Nystagmus is usually found in cerebellar abscess and rarely in cerebral.

Neurosyphilis

The brain and its meninges are involved in syphilitic gummatous affection. The inflammation usually starts in the chiasmal region and spreads over the base of the brain. Bilateral papillitis or papilledema or postneuritic optic atrophy is common. The third cranial nerve is involved in nearly 30 percent of cases, less frequently, the fifth and the sixth, and least frequently, the fourth cranial nerve may be affected.

Primary optic atrophy is found in about 20 percent cases of *tabes dorsalis*. Argyll Robertson pupil, concentric contraction of visual fields and internal and external ophthalmoplegia may occur. Locomotor ataxia is an important sign of the disease. It is predominantly seen in males, most frequently between 30 and 50 years of age.

Ocular signs and symptoms are more common in *general paralysis of the insane* or *paralytic dementia* seen in tertiary syphilis. The disease is now-a-days quite rare because of early diagnosis and treatment with penicillin. The general paralysis is usually accompanied by tabetic signs. Argyll Robertson pupil (50%), primary optic atrophy (8%) and external ocular muscle paresis due to the third cranial nerve involvement are common. Progressive paralysis and dementia are not rare.

DEMYELINATING DISEASES

Ocular symptoms and signs are found in almost all the demyelinating diseases.

Multiple Sclerosis

Multiple sclerosis produces patchy demyelination of the visual tract. It is more common in women than men. Ocular pain, diplopia, optic neuritis, central or centrocecal scotoma, nystagmus and paralysis of extraocular muscles are important ocular features. The symptoms appear suddenly but near complete recovery is not rare. Recurrences are common.

Neuromyelitis Optica (Devic's Disease)

Neuromyelitis optica is an inflammatory disease of the CNS that causes damage to the myelin sheath of nerves. It affects the optic nerve and spinal cord but the brain is typically normal. Sudden bilateral optic neuritis precedes the signs of myelitis in Devic's disease. Recovery is often partial and the course of the disease is highly variable.

Diffuse Sclerosis (Schilder's Disease)

Diffuse sclerosis is characterized by widespread demyelination of cerebral hemisphere occurring in young people. The destruction of visual centre and optic radiations causes cortical blindness. Optic neuritis or retrobulbar neuritis, ophthalmoplegia and nystagmus are common.

VASCULAR LESIONS

Intracranial Aneurysms

Intracranial aneurysms affecting the circle of Willis are of ophthalmic interest. The aneurysms may be congenital or due to an injury or arterial degeneration. They cause ocular symptoms in three ways.

1. By mechanical pressure on the underlying structures
2. Fleeting symptoms due to sudden increase in the size causing ptosis, diplopia and visual impairment, and recovery following the leakage, and
3. By sudden bursting, leading to an apoplectic attack associated with subarachnoid hemorrhage.

Vascular Occlusion

The occlusion of the *posterior cerebral artery* in the occipital cortex causes crossed homonymous hemianopia with sparing of the macula.

The occlusion of the *middle cerebral artery* affects the optic radiation and causes *visual agnosia,* where a person can perceive an object but has no meaningful associations to it (inability to recognize familiar faces or objects), with crossed homonymous field defects.

The occlusion of the *posterior inferior cerebellar artery* results in nystagmus and Horner's syndrome along with medullary signs.

The occlusion of the *anterior cerebral artery* causes unconsciousness, contralateral hemiplegia with turning of the eyes and the head towards the side of lesion. On recovery the eyes deviate towards the opposite side.

Hemorrhage

Subarachnoid hemorrhage usually occurs due to the rupture of an intracranial aneurysm, angioma or atheromatous artery. It is also found in patients of blood dyscrasias and following trauma. It causes sudden and severe headache associated with vomiting and dizziness. The patient becomes comatose and may even die. If the patient recovers from coma, moderate papilledema, retinal hemorrhages (subhyaloid), ocular motor nerve palsies, visual field defects and proptosis may be noticed. Blood in the cerebrospinal fluid may be seen.

Hemorrhage in the frontal motor cortex causes conjugate deviation of the eyes away from the side of lesion during the irritative stage which is reversed in the paretic stage.

Hemorrhage in the internal capsule causes conjugate deviation of the head and eyes to the same side of lesion with contralateral hemiplegia.

Pontine hemorrhage produces conjugate deviation of the eyes and head away from the side of lesion. It causes extremely constricted pupils and contralateral hemiplegia.

HEAD INJURIES

Concussion injuries often cause subdural hemorrhage and unconsciousness. Initially, the ipsilateral pupil is constricted; later owing to raised intracranial pressure the pupil dilates and does not react to light (*Hutchinson's pupil*). Similar changes occur in the contralateral pupil if the pressure continues to rise. The presence of dilated fixed pupils warrants cerebral decompression.

Fracture of the base of skull produces cranial nerve palsies. Ipsilateral facial paralysis of lower motor neuron type is most common. Sixth and third cranial nerves may also be involved. Presence of papilledema suggests hemorrhage into the nerve sheath. The pupillary reactions are inconsistent.

INTRACRANIAL TUMORS

Intracranial tumors frequently give generalized symptoms of raised intracranial pressure such as headache, nausea, vomiting, vertigo, convulsions, and alterations in pulse, respiration and blood pressure, and papilledema. Localizing signs—pupillary changes, specific field defects and ocular palsies—are seen due to pressure on or destruction of underlying or neighboring structures.

Severe papilledema is found with tumors of the midbrain, parieto-occipital region and cerebellum. Ventricular tumors cause moderate degree of papilledema. Nearly 50 percent of pontine tumors give rise to papilledema. Tumors of the medulla oblongata and the central white matter of the cerebral hemisphere do not cause papilledema. Meningioma of the olfactory groove induces ipsilateral pressure optic atrophy and contralateral papilledema due to increased intracranial pressure (*Foster-Kennedy syndrome*).

Temporal Lobe Tumors

Temporal lobe tumors are associated with papilledema in 50 percent of cases. They produce incongruous crossed upper quadrantanopia owing to pressure on the optic radiations. Visual hallucinations occur due to irritation of visuopsychic or circumstriate areas. Third and fifth cranial nerves may be involved due to downward pressure.

Parietal Lobe Tumors

The tumors of the parietal lobe give a crossed lower homonymous quadrantanopia due to pressure on the upper fibers of the optic radiations. Visual and auditory hallucinations, conjugate deviation of the eyes and optokinetic nystagmus may also occur.

Occipital Lobe Tumors

Occipital lobe tumors give essentially the visual symptoms. Crossed homonymous quadrantic or hemianopic field defects, possibly involving the fixation point are characteristic. Visual agnosia may be found.

Tumors of the Brainstem

The *tumors of the upper part of midbrain* (involving the colliculi and pineal gland) cause spasmodic contraction of the upper lid followed by ptosis and are associated with loss of the conjugate movements of the eyeball upwards and, sometimes downwards. Occasionally, an upper motor neuron facial paralysis and ipsilateral hemiplegia may develop.

The *tumors of the intermediate part of midbrain* (cerebral peduncles) cause ipsilateral third nerve palsy with contralateral hemiplegia (*Weber's syndrome*). When the red nucleus is involved, the ipsilateral third nerve palsy is associated with tremors and jerky movements (ataxia) of the contralateral side of the body (*Benedikt's syndrome*).

The *tumors of the pons* implicate the pyramidal tracts and involve the third, fifth, sixth, seventh and eighth cranial nerves. If the lesion is in the lower part of the pons, it causes ipsilateral sixth nerve palsy, ipsilateral facial palsy and contralateral hemiplegia (*Millard-Gubler's syndrome*). Tumors of the dorsal pons implicate the paramedian pontine reticular formation (the site for horizontal gaze center) and cause *Foville's syndrome* which is characterized by ipsilateral facial analgesia (fifth cranial nerve affection), sixth nerve palsy along with gaze palsy, facial weakness (seventh nerve involvement) and deafness (eighth nerve palsy).

Cerebellar Tumours

The *tumors of cerebellopontine angle* give rise to corneal anesthesia (due to the fifth cranial nerve involvement), hearing loss, sixth and seventh cranial nerves paresis and cerebellar symptoms. Cerebellar tumors also cause marked papilledema and nystagmus.

Chiasmal and Pituitary Tumors

The chiasmal tumors cause characteristic visual field defects—bitemporal hemianopia. Suprasellar tumor (craniopharyngioma) leads to papilledema. Primary optic atrophy and ocular motor nerve palsies are not uncommon. Acidophilic adenoma of the pituitary gland produces gigantism or acromegaly, while other pituitary adenomas give signs of hypopituitarism.

DISORDERS OF CRANIAL NERVES

Trigeminal Neuralgia (Paroxysmal Facial Pain)

Trigeminal neuralgia is characterized by excruciating pain on the lips, gums, cheek, chin and, rarely, in the distribution of the V cranial nerve. The cause of the pain is not assignable. Treatment includes oral carbamazepine 100-200 mg four times a day. When drug fails to relieve the pain surgery is indicated.

Bell's Palsy

Bell's palsy is the most common type of facial palsy. The pathogenesis of the palsy is unknown although an inflammation of the facial nerve producing pressure on the nerve as it leaves the skull within its bony canal is the most accepted hypothesis. The pain behind the ear may precede abrupt Bell's palsy. The facial palsy may also occur in Ramsay Hunt syndrome (Herpes zoster oticus), acoustic neuroma (an intracranial tumor of myelin-forming cells of the vestibulocochlear nerve commonly associated with neurofibromatosis), uveoparotid fever (uveitis, parotid gland enlargement and facial palsy), Lyme disease and leprosy. Bell's palsy is a diagnosis of exclusion. Systemic corticosteroids and a course of oral acyclovir are quite effective.

Ocular Motor Nerve Palsies

An isolated third nerve palsy may be caused by compression by an aneurysm of the posterior communicating artery at its junction with the internal carotid artery. It manifests as a total third nerve palsy.

An isolated sixth nerve palsy may occur due to compression by an aneurysm of the internal carotid artery in its intracavernous course.

Multiple ocular motor nerve palsies (III, IV and VI cranial nerves) can occur due to cavernous sinus lesions.

DEGENERATIVE DISEASES

Ocular Myopathy

Ocular myopathy or mitochondrial encephalomyelopathy is characterized by a progressive external ophthalmoplegia and weakness of muscles of the face, neck and limbs. Ptosis is often a presenting sign. Weakness of the extraocular muscles eventually develops into a complete external ophthalmoplegia which is accompanied with wasting of facial muscles. There is no specific treatment for ocular myopathy.

Wilson's Disease

Wilson's disease is a familial hepatolenticular degeneration characterized by tremors of head and limbs, generalized rigidity and hepatomegaly. A characteristic golden-brown ring in the periphery of cornea (Kayser-Fleischer ring) may be present. The ring represents the deposition of elemental copper in the region of Descemet's membrane. Typically, it appears first superiorly, then inferiorly, medially and finally laterally. It is visible on slit-lamp biomicroscopy. Treatment consists of avoiding intake of food rich in copper (nuts, chocolates, organ meat), oral chelation therapy with penicillamine or trientine (to remove copper from potentially toxic sites) and zinc salts (to block the intestinal absorption of copper).

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CHAPTER

29 Ocular Manifestations of Systemic Disorders

Nutritional deficiencies, infective diseases, metabolic disorders, parasitic infestations and skin diseases often involve the eye in varying degrees. A clinician should be conversant with ocular symptoms and signs of these disorders.

NUTRITIONAL DEFICIENCIES

A deficiency of vitamin A causes night-blindness, conjunctival xerosis, corneal xerosis, keratomalacia and xerophthalmia. Vitamin A is an important vitamin essential for maintaining the structure and function of the ocular surface. The ocular signs are accentuated in the presence of protein calorie malnutrition and secondary bacterial infections.

Deficiency of vitamin B_1 (thiamine) occurs primarily in malnourished chronic alcoholics causing external ophthalmoplegia, nystagmus, ptosis, nonreacting miotic pupils, retinal hemorrhages and optic neuropathy (as a part of Wernicke's encephalopathy or dry beriberi).

Deficiency of B_2 (riboflavin) produces photophobia, conjunctival irritation and corneal vascularization. Its deficiency is also implicated in cataractogenesis.

Deficiency of vitamin C may produce hemorrhages in the conjunctiva, retina and orbit. It also delays the wound healing.

Vitamin D deficiency may be associated with zonular cataract and papilledema.

INFECTIVE DISEASES

Bacillary infections such as typhoid fever causes infective retinitis, optic neuritis, metastatic choroiditis, corneal ulcer, lid abscess, and subconjunctival, vitreous and retinal hemorrhages. Brucellosis can lead to uveitis. Whooping cough gives a characteristic black eye (subconjunctival hemorrhages) in children.

The common ocular manifestations of tuberculosis, leprosy and syphilis include conjunctivitis, keratitis, uveitis, vitritis, chorioretinitis and optic neuritis.

Lyme disease causes erythema and lymphadenopathy as early manifestations. Neurological and cardiac complications may follow. Chronic arthritis of large joints and polyneuropathy are late complications. The ocular features of the disease include photophobia, pain, keratitis, uveitis and optic neuritis.

Viral infections frequently implicate the eye. Conjunctivitis, corneal ulcer, uveitis, optic neuritis and endophthalmitis may be found in measles. Mumps often leads to dacryoadenitis and sometimes, keratitis. Influenza may be associated with conjunctivitis and, rarely, optic neuritis. Dengue fever may cause keratitis, iridocyclitis and ophthalmoplegia. Rubella infection, during the first trimester of pregnancy, causes congenital cataract, microphthalmos and pigmentary retinopathy.

Fungi such as *Aspergillus*, *Candida*, *Fusarium* and *Mucor* are capable of causing keratomycosis, endophthalmitis and orbital cellulitis.

PARASITIC INFESTATIONS

Retinal hemorrhages, keratitis and optic neuritis may be found in malaria. Retinal hemorrhages are common in kala-azar and keratitis in trypanosomiasis. Toxoplasmosis causes a characteristic punched-out central retinochoroiditis (Table 29.1). *Ecchinococcus granulosus* may produce hydatid cyst in the eye and orbit. *Cysticercus cellulosae*, the larval stage of tapeworm, may induce intravitreal or retinal inflammatory reaction causing leukocoria, and subconjunctival, subretinal and intravitreal cysts formation. Ocular larva migrans is caused by *Toxocara canis*.

Table 29.1: Ocular features of parasitic diseases

Parasitic disease	*Ocular features*
Toxoplasmosis	Macular scarring, retinochoroiditis, vitritis
Toxocariasis	Vitritis, choroiditis, vitreoretinal granuloma
Cysticercosis	Subconjunctival, subretinal, and vitreal cysticercus cysts
Onchocerciasis Acanthamoeba	Sclerosing keratitis, uveitis, cataract keratitis

INBORN ERRORS OF METABOLISM

Inborn errors of metabolism are caused by enzyme disorders. They are usually genetically determined. These errors of metabolism may be classified as following:

1. *Carbohydrate metabolism disorders*
   Diabetes mellitus
   Galactosemia
   Glucose 6-phosphate dehydrogenase deficiency
2. *Amino acid metabolism disorders*
   Albinism
   Chédiak-Higashi disease
   Homocystinuria
   Marfan's syndrome
   Hyperlysinemia
   Cystinosis
   Phenylketonuria
   Hyperornithinemia
3. *Mucopolysaccharidoses (MPS)*
   I to VI types of MPS
4. *Lipid metabolism disorders*
   Refsum's syndrome
   Bassen-Kornzweig syndrome
   Hyperlipoproteinemia
5. *Sphingolipidoses*
   Farber's disease
   Niemann-Pick disease
   Gangliosidoses
   Gaucher's disease
   Fabry's disease
6. *Mineral metabolism disorders*
   Wilson's disease
   Hemochromatosis.

It is beyond the scope of this book to review all the clinical features of inborn errors of metabolism. However, it is interesting to note that the ocular signs of some of the inborn errors of metabolism are striking and the ophthalmologists are the first persons to clinch the diagnosis.

Carbohydrate metabolism disorders are often associated with cataract. Diabetes mellitus presents with a typical retinopathy.

Amino acid metabolism disorders may be accompanied with lens dislocation, pupillary abnormality, mental retardation, laxity of joints and bone dysplasia. Classical iris transillumination is found in albinism and scalloped border peripheral chorioretinal atrophy (gyrate atrophy) is associated with hyperornithinemia. Birefringent crystalline deposits in the corneal stroma may be present in cystinosis.

Corneal clouding, tapetoretinal degeneration, mental retardation and skeletal dysplasia are seen in mucopolysaccharidoses.

Lipid metabolism disorders may be associated with retinal pigmentary degeneration, fatty skin deposits and progressive neuropathy.

Sphingolipidoses may present with cherry-red spot at macula, infantile cortical degeneration and splenohepatomegaly. The whorl-like corneal dystrophy (*vortex keratopathy*), spoke-like capsular lens opacity and dermal angiokeratoma (tiny, painless papules appearing in any region of body) are found in Fabry's disease.

Mineral metabolism disorders often lead to pigment deposition in ocular tissues. Classical Kayser-Fleischer ring (copper deposition at the peripheral part of Descemet's membrane) in Wilson's disease and slaty blue discoloration of the peripapillary fundus in hemochromatosis (excessive accumulation of iron in the body due to improper metabolism) are commonly seen.

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CHAPTER

30 Community Ophthalmology

The modern medical science is often accused of laying an undue emphasis on the study of disease and neglect of positive health. Prevention is better than cure, but people rely on drug therapy more than observing the natural laws governing health.

The World Health Assembly has declared that health is a basic human right and suggested that the governments of various countries and the World Health Organization (WHO) to work for it. However, the objective of the WHO to provide health to all the people of the world seems a distant dream. In fact *health* is a state of complete physical, mental and social well-being and not merely an absence of disease or infirmity. This concept of health has also been adopted in ophthalmology and a new branch, community ophthalmology, has emerged to combat blindness. *Community ophthalmology* is defined as a discipline of medicine which utilizes the methodologies of public health, community medicine and clinical ophthalmology to promote ocular health and prevent blindness.

BLINDNESS

Definition

Generally, blindness implies inability to perceive light, but different definitions of it are in vogue. From the economic standpoint, if a person is unable to perform the work for which eyesight is essential, he is referred as blind. An acceptable definition of blindness is proposed by the WHO for the purpose of collecting uniform statistics. A person having a visual acuity of less than 3/60 or 10/200 with correcting glasses in the better eye in day light is defined as *blind*. A concentric contraction of visual field to an average radius of 10 degrees is considered equally disabling .

The vision loss may range from mild to profound. In almost all surveys the WHO has adopted two levels of visual loss, *low vision*: < 20/60, < 0.3 or < 6/18 and *blindness*: < 2/400, <0.05 or < 3/60. Table 30.1 shows the various degrees of visual loss.

Table. 30.1: Range of visual loss and blindness

Grade of visual loss		*Visual acuity in different notations*		
1	Mild	20/50	0.4	6/18
2	Moderate	20/125	0.16	6/36
3	Severe	20/400	0.05	3/60
4	Profound	20/630	0.032	2/60
5	Blind	00	No LP	No LP

LP: Light Perception
The WHO has categorized Grade 1 and 2 as visual impairment while 3, 4 and 5 are referred as blind.

Global Blindness

The problem of blindness is worldwide. It occurs in the developed as well as developing countries. According to the WHO estimate nearly 37 million people worldwide are blind, 124 million people have low vision and 153 million people have visual impairment due to uncorrected refractive errors. Nearly 75-80% of world's blind reside in Africa and the Asian subcontinent. The number of blind is increasing every year by 1-2 million with the rise in population and improved average life expectancy.

Economical Impact

The impact of blindness on global economy is striking according to the data from world development report and global blindness (Table 30.2). The total Gross National Product (GNP) lost as a result of both childhood and adult blindness combined ranges from $ 167518 million to $ 243938 million.

Economical burden of blindness in India was estimated in the year 1997. Direct and indirect economic loss due to adult and childhood blindness was calculated. It comes to approximately Rs. 159 billion (US$ 4.4 billion). The cost of treating all cases of cataract blindness only in India is Rs. 5.3 billion.

Causes of Blindness

The WHO has listed six major causes of blindness (Table 30.3). Cataract, glaucoma and diabetic retinopathy occur worldwide while trachoma, vitamin A deficiency and onchocerciasis occur regionally.

Table 30.3: Prevalence of worldwide blindness on the basis of etiology

Etiology	*% of blindness*
Cataract	43
Glaucoma	15
Trachoma	11
Vitamin A deficiency	6
Onchocerciasis	1
Others (DR, AMD, optic neuropathy etc.)	24

The causes of blindness in developed countries are quite different from that in developing or underdeveloped ones. In the former, glaucoma, myopia, developmental anomalies, macular degeneration, diabetic retinopathy and cataract are the major causes of blindness, while cataract, infectious diseases of the eye (trachoma and allied conditions), malnutrition, injuries and glaucoma are common causes in developing or underdeveloped countries. Onchocerciasis occurs endemically in the West equatorial Africa and parts of Central and South America and renders millions blind.

Age and Blindness

The age is considered as a major risk factor for visual impairment. The rate of visual impairment increases with the increase in the age. The rate of blindness in the age group 50-59 years is 4.2%, the figure reaches 15.8% after the age of 70 year. The causes of blindness in childhood and adults are described below.

Table 30.2: Average GNP loss due to blindness

Population (million)	*GNP per capita in US $*	*Prevalence of blindness (%)*	*No. of blind (million)*	*Total working years lost due to blindness*	*GNP per capita loss due to blindness in US $*
5500	4420	0.7	Adult 36.5 Children 1.4	6-10 0-50	4420-5940 4420-19377

Childhood Blindness

There are as many as 1.5 million blind children in the world. It is estimated that a large number of the blind children live in Asia and Africa. The prevalence of childhood blindness varies from 9/100000 (UK and USA) to 100/100000 (Kenya). Table 30.4 shows the global prevalence of childhood blindness.

The causes of childhood blindness are starkly different in developed and developing countries (Table 30.5).

Congenital cataract, congenital glaucoma and other developmental anomalies can cause blindness in newborn. Vitamin A deficiency, refractive errors and amblyopia may cause severe visual loss in preschool and school-going children (Table 30.6).

Cataract: Approximately 200000 children worldwide are blind from cataract. Cataract in childhood may occur due to either genetic influence or intrauterine infections.

Table 30.4: Global distribution of childhood blindness

Region	*Prevalence per 1000 children*	*Estimated No. of blind children*
Africa	1.1	264000
Asia	0.9	1080000
South and Central America	0.6	78000
Europe, Japan and USA	0.3	72000
Total		1494000

Table 30.5: Causes of childhood blindness in developed and developing countries

Developed countries	*Developing countries*
Retinopathy of prematurity	Corneal scarring
Congenital cataract	Trachoma
Retinal dystrophies	Vitamin A deficiency
Congenital malformations	Cataract
Nystagmus	Ophthalmia neonatorum
	Congenital anomalies

Table 30.6: Common causes of blindness in newborns, preschool and school-going children

Newborns	*Preschool and school children*
Congenital cataract	Refractive errors
Congenital glaucoma	Vitamin A deficiency
Developmental anomalies	Strabismus
Nystagmus	Amblyopia
Strabismus	Trauma

Corneal scarring: Corneal scarring is the second most common cause of childhood blindness. Corneal scar may result from trachoma, trauma, microbial keratitis and ophthalmia neonatorum. The incidence of ophthalmia neonatorum may be as high as 10% of all births in East Africa. Trachoma and microbial keratitis cause severe scarring in adults.

Retinopathy of prematurity (ROP): ROP is a vasoproliferative disorder of the retina occurring in premature children. It is worldwide in distribution and one of the important causes of childhood blindness. Prematurity and low birth weight (less than 32 weeks gestational age and birth weight about 1000 g) are most important risk factors for the development of ROP. In a recent study, 49% of infants with approximately 1251 g weight developed ROP, of these nearly 7% reached threshold level. Prevalence of visual impairment caused by ROP greatly varies in most retrospective studies.

Vitamin A deficiency: Vitamin A deficiency is a major cause of both blindness and increased mortality among preschool children in developing and underdeveloped countries. The results of various surveys indicate that 5-10 million children suffer from xerophthalmia every year, of them 0.5 million develop serious visual impairment. Vitamin A deficiency is highly prevalent in Africa and South-East Asia. It has moderate prevalence in some countries of Central and South America. The prevalence of vitamin A deficiency is high

among the children of poor socio-economical stata. Poverty, poor diet, worm-infestation and bowl disorders are considered important risk factors for xerophthalmia.

Amblyopia: Amblyopia can result in monocular or binocular vision loss. It more frequently causes monocular visual loss. The incidence and prevalence of amblyopia vary from 1 to 2.5% in children. Amblyopia is almost always associated with the presence of anisometropia or strabismus. It can also result from visual deprivation (axial corneal opacity or cataract).

Ocular trauma: Ocular trauma is a cause of monocular blindness globally. It is estimated that 5% of total blindness is directly related to ocular trauma. Accidents or injuries can cause mortality, morbidity and disability. The WHO program for the prevention of blindness reports that 55 million eye injuries occur worldwide each year. Approximately 1.6 million become blind following ocular trauma. Additionally 19 million people suffer from visual impairment. Ocular injuries are seen more frequently in farmers in rural areas.

Serious penetrating injuries may occur in children during unsupervized play. Ocular trauma can be sustained during sports. Badminton and tennis players may suffer blunt trauma from shuttlecock and tennis ball respectively, while contusion injuries are common in boxers resulting in retinal damage. Chemical and blast injuries may occur during certain festivals. These injuries often involve both eyes resulting in blindness. In spite of seat belt legislation, eye injuries in road traffic accidents are common.

Trachoma: Trachoma is a leading cause of blindness. According to the WHO estimate trachoma causes 15% of blindness and affects about 150 million people and renders 6 million blind. The distribution of trachoma is heterogeneous. It is endemic in the Middle East, certain countries of Africa and South-East Asia, Central Australia and North Eastern Brazil. In trachoma endemic zones more than 50% of children suffer from active trachoma and majority of adults have varying degree of scarring. Scarring of conjunctiva may cause trichiasis, entropion and corneal scar. Trachoma Control Programs are introduced in areas where prevalence of active trachoma is higher than 10% in children aged 10 years.

Blindness in Adults

The WHO estimates that 26-34 million adults are blind in the world. The prevalence of blindness in adults differs in various eye disease surveys mainly due to variations in the definition of blindness and methodology. The prevalence rate of profound visual impairment (< 3/60 of visual acuity or < 5 degree of visual field) in published surveys ranges from 0.16 to 2.23%. The common causes of visual impairment in adults are described below.

Cataract: The most important cause of visual impairment in adults is cataract. Nearly 17 million people are blind from cataract. It is estimated that by the year 2020 40 million will be blind from cataract worldwide. Majority of cataract is age-related and occurs more frequently after the age of 60 years. In some tropical countries cataract develops at an early age and progresses rapidly. Data obtained from surveys indicate that nuclear and cortical types of cataract are more common than posterior subcapsular. Diabetic patients are at a higher risk for cataract formation especially the cortical. Cigarette smokers have an increased risk for development of nuclear cataract. Presently, no therapeutic agent is known to prevent the onset or progression of cataract.

Glaucoma: Glaucoma is a major cause of blindness in old age (> 60 years) in developed and developing countries. The prevalence of primary

open-angle glaucoma (POAG) ranges between 1 and 3% in white and Asian population, but in black population it ranges between 3 and 9%. The visual impairment caused by POAG is 6 times higher among the black compared to the white. Glaucoma is responsible for about 15% of all blindness. It is estimated that nearly 600000 people per year go blind from glaucoma globally. Some reports suggest that by the year 2020, glaucoma will be the leading cause of blindness worldwide followed by cataract and trachoma. Glaucoma will be responsible for nearly 14% of estimated 54 million blind people over the age of 60 years. The POAG is more common worldwide but primary angle-closure glaucoma (PACG) has higher prevalence than POAG in the East and South-East Asia. The prevalence of PACG in the population aged 40 years and older varies greatly in various ethnic groups: Europeans, 0.1-0.6%, Asians, 0.4-1.4% and mixed population of Africa, 2.3%. A survey conducted in the state of Andhra Pradesh reported 1.1% incidence of PACG. Early detection, proper treatment and periodic follow-up are essential for the prevention of blindness from glaucoma.

Microbial keratitis: Superficial or deep microbial keratitis is a leading cause of corneal blindness. It is responsible for at least 1.5 million new cases of monocular visual impairment worldwide every year. Microbial keratitis mostly affects farm-workers and poor laborers in the developing countries causing blinding sequelae. The incidence of corneal ulceration in Southern India was reported as 113 per 10000, which is 10 times higher than that in US.

Human immunodeficiency virus/Acquired immune deficiency syndrome (HIV/AIDS): The WHO and the Joint United Nation Program on HIV/AIDS reported that approximately 40 million HIV-positive people are presently living across the globe. Majority of these people live in developing countries. Nearly 75% of HIV-positive patients develop opportunistic infections such as CMV retinitis, HZO, Kaposi sarcoma and HIV retinopathy resulting in serious visual loss.

Diabetic retinopathy (DR): Diabetic retinopathy is a leading cause of blindness in adults and aged persons. The duration of diabetes is the most important risk factor for the development of retinopathy. DR is uncommon in patients with duration of less than 10 years of diabetes. After a duration of 20 years of Type I diabetes, proliferative diabetic retinopathy (PDR) develops in 30 to 40% of patients. The occurrence of clinically significant macular edema (CSME) also increases with the duration of the disease. Nearly 25% of patients with diabetes of more than 25 years develop CSME. An intensive glycemic control in patients with diabetes reduces the risk of development of DR as well as retards the progression of DR. Panretinal photocoagulation is an important modality in the management of PDR. It significantly reduces the blindness from DR as compared to those without panretinal photocoagulation according to Diabetic Retinopathy Study. The Early Treatment Diabetic Retinopathy Study (ETDRS) reported that focal photocoagulation for CSME reduces the risk of visual loss.

Onchocerciasis: Onchocerciasis is caused by a filarial worm *Onchocerca volvulus*. The worm has human host and an intermediate host, the blackfly of genus *Simulium*. Onchocerciasis is endemic in some areas of Africa and South and Central America. In these areas nearly 60% of the population may be affected by the disease and the rate of visual impairment ranges from 5 to 10%. However, the rate of blindness is found to be low (1.5%) in West African forest, though the prevalence of onchocerciasis is more or less the same. The disease predominantly affects the males above the age of 40 years. Corneal scarring,

glaucoma, chorioretinitis and optic atrophy may cause blindness in onchocerciasis. The Onchocerciasis Control Program (OCP) is focused on control of the vector and different insecticides are spread in rotation to avoid any chance of development of resistance. The results of OCP are very good and prevalence of the disease has significantly come down. In the community treatment of onchocerciasis, ivermectin is given in a dose of 150 microgram per kilogram body weight, once a year. The drug should not be given in children below the age of 5 years and in pregnant women. This treatment program has high compliance and provides encouraging results.

BLINDNESS IN INDIA

The sample survey (1975) conducted in India provides a comprehensive data on the causes of blindness. Cataract was the main cause accountable for 55 percent of blindness. It was followed by ocular infections (20%). Malnutrition was responsible for 2 percent, smallpox 3 percent, injuries 1.25 percent and glaucoma 0.5 percent of total blindness. Congenital disorders, uveitis, retinal detachment, tumors, diabetes, hypertension and neurological disorders contributed for the remaining 18 to 25 percent of blindness.

A recheck survey was conducted (1986-89) under the aegis of the World Health Organization and the National Program for Control of Blindness (WHO-NPCB). It revealed that cataract is a major cause of blindness accounting for 80.1 percent of blindness in India (Table 30.7).

A report of Eye Disease Study by the National Program for Control of Blindness in India (1992) stated that cataract caused 60.8% blindness but a regional survey conducted by Andhra Pradesh Eye Disease Study (APEDS) reported that cataract was responsible for only 34.3% of blindness (Table 30.8).

Table 30.7: Causes of blindness in India (1986-89)

Disease	*Blindness%*
Cataract	80.10
Refractive errors	7.35
Aphakic blindness	4.67
Glaucoma	1.00
Corneal opacity	1.52
Trachoma	0.39
Others	4.25

Table 30.8: Causes of blindness in EDSNPCB and APEDS surveys in India

Cause	*EDSNPCB survey (% of blindness, vision < 3/60)*	*APEDS survey (% of blindness, vision < 3/60 or < 10° visual field)*
Cataract	60.8	34.3
Retinal diseases	6.3	22.4
Corneal diseases	23.6	20.1
Glaucoma	0	15.2
Optic atrophy	7.4	6.4
Trauma	1.9	1.6

EDSNPCB: Eye Disease Study National Program for Control of Blindness, APEDS: Andra Pradesh Eye Disease Study

Childhood Blindness in India

There are so far no reliable data on the prevalence of childhood blindness from India. A rough estimate indicates that there are 200000 (+ 50000) blind children in India which seems to be an underestimate. Common causes of childhood blindness in India include corneal scar (26%), congenital anomalies of the globe (25%), diseases of the retina (21%) and cataract (12%). The corneal scarring is mainly due to vitamin A deficiency, trachoma and conjunctivitis.

The number of people rendered blind due to corneal disorders is small as compared to cataract, but the patients are mostly children or adolescents, the most productive members of the society, unlike elderly cataract patients. The corneal blindness can

be prevented. Children at high-risk of vitamin A deficiency should be treated with vitamin A capsule 200000 IU on day 1, 2 and 14 (the dose for infants is just the half). Immunization against measles and early control of diarrhea are important strategies to prevent corneal xerosis.

Early management of trachoma, conjunctivitis and corneal ulcer prevents corneal damage. Although in the past three decades, the blindness due to trachoma and conjunctivitis is substantially reduced, still there are more than 0.5 million blind owing to corneal pathology. The vision can be restored in these blind persons by corneal grafting. Presently there are about 505 eye banks in the country performing nearly 12975 keratoplasties each year. Many corneal blind people continue to wait for the want of donor cornea. Therefore, eye donation campaign is an urgent need. Both the government and voluntary agencies have to promote awareness in the society regarding eye donation.

Besides the above mentioned components of community ophthalmology, school eye health programs and occupational eye health services help in controlling the childhood blindness from amblyopia and trauma respectively.

Adulthood Blindness in India

High myopia, trauma, abiotrophic defects, uveitis and neurological disorders are responsible for blindness in adult life. The ocular hazards can happen both at work and at play leading to serious visual loss. Industrial eye accidents resulting in blindness occur in every country. The number is far greater in developing countries like India. Road traffic accidents and sports injuries also contribute to blindness. Recurrent uveitis reduces the visual acuity significantly. Multiple sclerosis and brain tumors lead to severe visual handicap.

Old Age Blindness in India

Blindness in elderly persons occurs mainly due to cataract, glaucoma, diabetic retinopathy, age-related macular degeneration and retinal vascular disorders. In India nearly 3.8 million people become blind from cataract each year. Senile cataract develops at a relatively younger age in India than in developed countries. With the introduction of low cost intraocular lens implantation technology in India, the restoration of good quality vision in cataract patient is feasible. However, due to paucity of resources, especially ophthalmic surgeons, a significant percentage of cataract patient remains unoperated resulting in a huge backlog. Many of these eyes are lost due to dislocation of the lens and secondary glaucoma, or spoiled by quacks.

Second only to cataract, as a cause of bilateral blindness, is glaucoma. The cataract blindness is reversible but the blindness caused by glaucoma is irreversible. The early diagnosis and management of glaucoma is a difficult issue. The coexistence of glaucoma and senile cataract may pose diagnostic problem and many eyes may be lost while waiting for cataract extraction.

Diabetes mellitus is an important cause of blindness in advanced age group. Diabetic retinopathy accounts for 5 to 10 percent of registered blind in USA and UK. Perhaps the percentage of blindness due to diabetes in India is much higher because of poor control of the disease.

In recent years, there has been a considerable increase in the longevity of people. It has resulted in more and more cases of age-related macular degeneration.

Factors Influencing the Prevalence of Blindness in India

Besides age, other factors which influence the rate of blindness include sex, ethnicity and availability of health care services. The prevalence of blindness is higher in women than in men mainly due to their preoccupation in household activities and relatively lower level of health consciousness. Moreover, they are more susceptible to infections during pregnancy.

The blindness is more prevalent in certain ethnic groups probably due to their customs and genetic trait. Severe visual loss is more common in people coming from a lower socioeconomic status as found in most population based surveys. The adverse environmental factors and non-availability or nonaffordability of the health care services lead to a higher prevalence of blindness as well as higher mortality and morbidity rates in poor people.

Besides poverty, low standards of personal and environmental hygiene, illiteracy, ignorance, superstition, scarcity of water and medical services beyond the reach of the poor contribute to the higher prevalence of blindness in under-privileged community. The blindness caused by infectious diseases in India has considerably declined of late. Blindness due to ophthalmia neonatorum and retinopathy of prematurity has reduced significantly owing to early diagnosis and proper treatment.

CONTROL OF BLINDNESS

Vision 2020

Approximately 80% of global blindness is avoidable or preventable. The WHO and the International Agency for Prevention of Blindness (IAPB) are working to reduce avoidable blindness worldwide. The IAPB has encouraged the establishment of National Society for Prevention of Blindness in different countries to implement the WHO health care strategies against blinding diseases. The WHO and IAPB in the year 1999 have jointly launched vision 2020, Right to Sight, a project to combat the gigantic problem of blindness in the world and to eliminate the avoidable blindness by the year 2020.

The vision 2020 is a global campaign supported by 12 task force members (non-governmental organizations) and 8 supporting members. Under this program about US$ 80 million per year is being spent on the prevention of blindness.

The main objective of Vision 2020 is to eradicate the avoidable blindness in order to give all people in the world, particularly the millions needlessly blind, right to sight. The project is targeted to attain the best possible vision for all people thereby improving their quality of life. This can be achieved through the establishment of a sustainable comprehensive eye care system as an integral part of every national health system. The 59th World Health Assembly in 2006 has given substantial support to this global initiative for prevention of blindness. It has urged its members to set up national vision 2020 plans and provide support to the project by mobilizing domestic funds. Vision 2020 has targeted the following 5 avoidable eye diseases:

1. Cataract
2. Trachoma
3. Onchocerciasis
4. Childhood blindness, and
5. Refractive errors and low vision.

The control of these diseases seems possible by adopting the following strategies:

1. Cost-effective disease prevention and control
2. Human resource development and training of personnel
3. Infrastructure strengthening
4. Use of appropriate and affordable technology for eye care delivery, and
5. Mobilization of resources.

The strategy of Vision 2020 is built upon a foundation of community participation.

Disease Prevention and Control

Cataract: Cataract is the foremost cause of blindness with an estimated backlog of 16-20 million unoperated cases. There is an urgent need to increase the number of cataract surgeries. It is envisaged that in the year 2010, 20 million cataract operations should be performed to check the backlog, and in the year 2020, the target should be 32 million.

Trachoma: Trachoma is responsible for nearly 5.6 million blind, and about 146 million active cases worldwide need the treatment. The SAFE (surgery, antibiotic treatment, facial cleanliness and environmental improvement) strategy is being introduced in 46 endemic countries. The WHO Alliance for the Global Elimination of Trachoma is established to control the disease. The job of this global initiative includes 5 million trichiasis operations and treatment of at least 60 million patients with active trachoma by 2010. It is expected that the blindness due to trachoma should be eliminated by the end of year 2020.

Onchocerciasis: Onchocerciasis Control Program is gaining popularity in endemic zones. It is expected that community-directed treatment with annual doses of ivermectin may probably eliminate this blinding disease from the endemic areas of Africa and Latin America.

Childhood blindness: There are estimated 1.5 million blind children in the world of them 1 million live in Asia and 300000 in Africa. There are an estimated 500000 children going blind each year. The aim of the project is to eliminate the avoidable causes of childhood blindness.

Refractive errors and low vision: Refractive errors are on priority of vision 2020 project as they cause worldwide visual disability. A global initiative is needed to correct the refractive errors by spectacles to prevent amblyopia. Patients with low vision need low vision devices. Refraction and dispensing of glasses are integrated with primary health care and school eye health programs.

Human Resource Development

There is a need to train more ophthalmologists in developing countries. Presently the ratio of ophthalmologist/population ranges from 1:500000 to 1:200000. The human resources development should bring it to 1:50000 to render better eye care services in developing countries. Additionally, the number of ophthalmic assistants and nurses should also be strengthened. The global initiative will train primary health care workers for eye care, school teachers for visual screening of school children, refractionist, managers for national and regional program, and also equipment technicians. It is envisaged that there should be 100% training in basic eye care in medical schools by the year 2020.

Infrastructure and Appropriate Technology Development

Strengthening of the existing eye care infrastructure or the infrastructure development is an essential component of the global initiative which ensures the availability of refraction facilities, basic eye medicines and eye beds for at least 90% of the population by the year 2020. With improved resources and medical manpower, a permanent infrastructure of eye care can be created. It will provide basic as well as specialized eye services to all the needy patients.

Appropriate technology will manufacture locally essential good quality and cost-effective tools of eye examination, instruments for cataract and trichiasis surgeries, and optical devices (eye glasses, magnifiers, etc.).

Mobilization of Resources

The resources are mobilized for the universal coverage and accessibility of services for the preservation of vision and restoration of sight.

BLINDNESS CONTROL PROGRAM IN INDIA

Public health strategies are capable of controlling the preventable blindness in India which is nearly

80 percent of total blindness. Blindness can be controlled by adopting the following strategies:

Health Education Program

Health education program is an important preventive measure. It can be carried out in schools, fairs and community gatherings. It generates awareness among the people about the importance of balanced nutrition, proper personal hygiene and clean environment, and it dispels ignorance and superstition. It creates health consciousness among the people and the population becomes interested in the eye care activities.

Training Program

In India the ratio of the number of ophthalmologists and paramedical personnel engaged in ophthalmic service to the population is low, and thus no viable ophthalmic service infrastructure can be created to meet the demands of existing eye patients. It is, therefore, necessary to train a large number of eye surgeons and paramedical personnel to cater the needs of the population.

Disease Priority

All eye diseases cannot be eradicated with the existing resources in terms of manpower and money. Hence, major blinding eye diseases have to be tackled on the priority basis. To identify such diseases a sample survey or screening project has to be undertaken in a community. Vitamin A-Protein Prophylaxis and Trachoma Control Projects have been successfully launched in India for the control of keratomalacia and trachoma respectively.

National Program for Control of Blindness

The World Bank and a few developed countries are providing financial assistance for the control of blindness. After the success of Trachoma Control Project and Smallpox Eradication Program, the Government of India has launched the National Program for Control of Blindness with an aim to reduce the prevalence of blindness from 1.49 to 0.3 percent. Since 80 percent of the Indian population resides in rural area, a three-tier eye care approach has been adopted.

Primary Eye Care

The primary eye care facilities have to be provided at every primary health center and subcenter. The staff posted at the center is trained in the methodology of prevention of blindness. The ophthalmic assistant of the center disseminates ocular health education, provides guidance to eye patients and refers them to an ophthalmologist.

Secondary Eye Care

The secondary eye care is provided by the ophthalmologists of the district or subdivisional hospitals. To facilitate proper eye care services, trained medical and paramedical staffs are posted and basic ophthalmic equipments are provided to the hospitals. The staff actively participates in the conduction of eye camps.

District Blindness Control Society has been formed in each district to coordinate the activities of the government as well as voluntary organizations engaged in the control of blindness in the district.

Tertiary Eye Care

The tertiary eye care services are available in the department of ophthalmology of all medical colleges and regional institutes of ophthalmology. These institutions provide not only specialized eye care (including corneal grafting and vitreo-retinal surgery) but also train ophthalmic surgeons (manpower development) to work at the district and subdivisional hospitals.

Apex Center

To supervise, guide and coordinate the activities of NPCB at all levels, an apex center is established at Dr Rajendra Prasad Center of Ophthalmic Sciences, New Delhi. The director of the center works as an ophthalmic advisor to the Government of India. The center provides advanced treatment to the referred eye patients and imparts training to postgraduates in ophthalmology.

Besides Rajendra Prasad Center, Aravind Eye Hospital, Madurai, Sankara Nethralaya, Chennai and LV Prasad Eye Institute, Hyderabad are other ophthalmic centers of excellence providing modern eye treatment facilities and comprehensive advanced training in subspecialities of ophthalmology. Many international agencies are helping these institutions in their training as well as community oriented programs.

Mobile Surgical Units

Establishment of mobile surgical units or eye camp services is a need based approach. It brings the service almost to the door of the patient's home. The eye camp service is not meant for the cataract operation only but provides a total eye care, and seems to be the best solution to the problem of blindness for the time being until a permanent effective infrastructure is developed.

To combat the cataract blindness in India the eye camp approach is adopted. The National Society for Prevention of Blindness has issued strict guidelines for the conduction of eye camp with a view to minimize the complications. The Madurai model of eye camp is quite safe wherein the cataract patients are brought to the main or satellite eye hospitals and quality eye surgery is performed. The patients are transported back to their native villages after a short follow-up.

In many eye camps, good quality surgery is made useless by providing wrong spectacles. The need for a postoperative proper refractive correction cannot be overemphasized inspite of the fact that intraocular lens implantation surgery in eye camps restores quality vision in most of the patients.

Vision 2020 in India

A Draft Plan of Action for Vision 2020 in India was approved in principle by Ministry of Health and Welfare in 2002 as a document of future planning of NPCB. Seven ocular diseases have been targeted: cataract, childhood blindness, refractive errors and low vision, corneal blindness, diabetic retinopathy, glaucoma and trachoma. The Vision 2020 program in India has following strategies to combat blindness:

1. Infrastructure and support for primary eye care
2. Human resource development and training
3. Models for service delivery and community participation.

Infrastructure Support for Primary Eye Care

Vision centers are established for primary eye care (PEC). Each vision center covers a population of 50000 in rural area and manned by Middle Level Ophthalmic Personnel (MLOP), dedicated paramedics and nurses. The primary eye care includes identification and referrals of external eye diseases, vision testing and prescription and dispensing of eye glasses, school eye screening program, eye health education, training of volunteers and identification and referral of patients with cataract and glaucoma to service centers.

Human Resource Development and Training

Emphasis is given on the subject of ophthalmology in undergraduate medical eduction to lay a sound foundation and, even after postgraduation, there must be continued professional improvement through continuing medical education and fellowship courses in subspecialities in ophthalmology. The desirable ratio of Ophthalmologist: MLOP is 1:3 or 1:4.

Models of Service Delivery and Community Participation

The services can be delivered through Government and NGO infrastructures, primary health centers, community based rehabilitation program and vision centers. The service delivery models should have high standard, affordable, feasible, and must involve the community. The community participation is encouraged in school eye screening, cataract identification, screening for refractive errors, diabetes and glaucoma, and follow-up and referrals.

Rehabilitation

Rehabilitation of the blind is an important aspect of the blindness control program. Rehabilitation is of two types—medical rehabilitation and social rehabilitation.

Medical rehabilitation can be achieved by performing corneal grafting in a blind who has lost the vision due to corneal scarring. Persons with macular dystrophy or high myopia can be helped by fitting low vision aids. Blindness due to cataract is curable and patients generally return to work after the optical correction following surgery.

Incurable blind people (due to glaucoma or neurological disorders) have to be socially rehabilitated and trained in such a way so that they can earn their livelihood. There are a few centers which are active in providing social rehabilitation to the blind. During vocational training the blind gets instructions on communication and learns how to walk on busy roads. They are also provided educational facilities either through Braille or through the recorded texts. Recently, vocational training facilities for the blind have immensely increased. They are trained as telephone operators, machine operators, computer programers, typists and pianists. The rehabilitation of all the blind seems a remote possibility. The International Agency for the Prevention of Blindness and other agencies are engaged in solving the problem of rehabilitation of blind by creating additional blind schools, blind homes and blind welfare organizations.

Incurably blind needs security from family, society and government. Their urge for economic or social benefits like special quota employment, pension and free or concessional travel is justified. Many of the developing countries are providing budgetary provisions to fulfil some of the demands of these people.

Since the number of blind schools and blind homes in India are few, the focus is on community-based rehabilitation. Attempts are ongoing to make a blind person socially acceptable and a productive member of the family.

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APPENDIX
OPHTHALMIC INSTRUMENTS

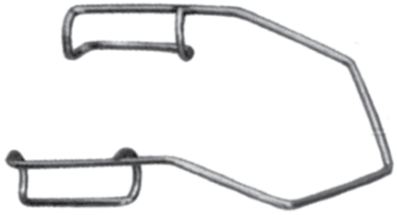
The quality of the ophthalmic instruments has markedly improved of late. Many new instruments have been developed to perform microsurgical procedures. Earlier instruments were made of stainless steel but now for non-cutting instruments titanium is used which is light in weight and resists corrosion. The steel is still used for most cutting instruments as it maintains the sharp edge for a long time. Many ophthalmic surgeons prefer disposable blades, needles, sutures, canulas and cutting instruments. Common instruments used in the ocular surgery are described below.

1. Eye Speculums

The eye speculum is of two types—universal and guarded. The universal speculum can be used for either eye. It consists of a spring and two limbs to keep the eyelids apart. The guarded speculum is separate for the right and the left eye. The guards keep the eyelashes away from the field of operation. However, the operative space becomes less. Eye speculums are used to separate the eyelids during surgery on the eyeball.

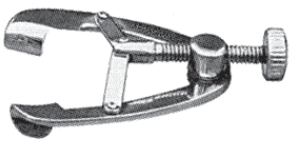
1.1. Barraquer's Wire Speculum

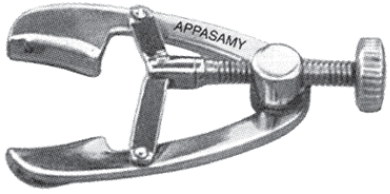
It is a universal type of eye speculum made up of wire to keep the lids apart. As it has a very light weight, it exerts minimal pressure on the eyeball.



1.2. Sauer's Infant Speculum

It is used for both premature and mature infants. The speculum has guards to retract the lids and eyelashes.





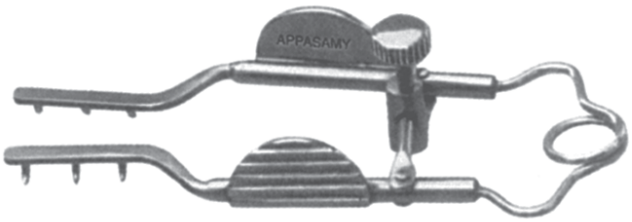
1.3. Clark's Speculum

It is a multipurpose speculum having a spring mechanism.

2. Retractors

2.1. Müller's Lacrimal Retractor

The retractor is made up of two limbs with a screw to fix the limbs in the retracted position. Each limb contains three curved pins for engaging the edges of skin incision. It is used for retracting the skin during dacryocystectomy and dacryocystorhinostomy operations.



2.2. Stevenson's Lacrimal Sac Retractor

The retractor has blunt prongs. It causes less trauma to the tissue. It is a self-retaining retractor.

2.3. Cat's Paw Retractor

It is a fork-like instrument. The terminals of the forks are bent downwards. Blunt and sharp retractors are available. The blunt one is used to retract the sac while the sharp is used to retract the skin and the ligament during sac surgery.

2.4. Dastoor's Iris Retractor

It is a delicate instrument used for retraction of the iris during phacoemulsification and cryoextraction of the lens during intracapsular cataract extraction (ICCE).

2.5. Desmarres' Retractor

It is a saddle-shaped instrument folded on itself at the end of a metal handle. It has multiple uses such as to examine the eyeball when there is marked blepharospasm, examination of the eyeball in children, removal of corneal or corneoscleral sutures and to obtain double eversion of the upper lid for examining the fornix.

2.6 Schepen's Orbital Retractor

Schepen's retractor has a notched blade and a handle. The forked retractor is used in retinal reattachment surgery (scleral buckling).

3. Foreign Body Spuds

3.1. Ellis Foreign Body Spud

It has a very thin, rounded and curved tip used for removal of a foreign body from the cornea.

3.2. Foreign Body Spud and Gouge

It is used for removal of superficial as well as deep foreign bodies from the cornea.

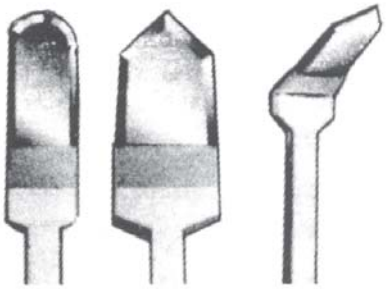
4. Knives

4.1. von Graefe's Cataract Knife

The knife has a long, narrow, thin and straight blade with a sharp tip and a fine sharp cutting edge. The knife was frequently used to make *ab interno* sclerocorneal section during intracapsular cataract surgery as well as for making section for iridectomy for glaucoma or optical purpose.

4.2. Diamond Knives

They are extremely sharp diamond-tipped knives. The tip can be straight or angulated.
The diamond knives are used to make corneal incisions in radial keratotomy, and corneal or corneoscleral section during an intraocular surgery.



4.3. Ziegler's Knife

It is a fine hook-shaped knife with a sharp pointed tip. It is used to incise the after cataract and perform capsulotomy. Ziegler's knife may be used to incise the iris in updrawn pupil after cataract surgery. It was earlier used for discission operation.

4.4. Tooke's Knife

It has a short flat blade with semicircular cutting edges, bevelled on both the surfaces like a chisel. It is used to expose the corneoscleral junction during cataract surgery, and to separate scleral lamellae during trabeculectomy or partial scleral resection.

4.5. Bard-Parker Knife

It consists of two parts: a flat or round handle and a number 11 or 15 blade. It is widely used in the ocular surgery such as chalazia, cataract and glaucoma surgery.

5. Blades and Keratomes

5.1. Super Sharp Blade

It is a very sharp blade used for making a side port incision.

5.2. Tunnel Blade

It is relatively blunt crescent-shaped blade used for making scleral tunnel in phacoemulsifiaction surgery and small incision cataract surgery (SICS).

5.3 Stiletto Blade

It is a lancet-shaped, 20 gauge, 1 mm wide blade attached to a handle. The micro vitreoretinal (MVR) blade is used for giving a stab incision in the cornea or the sclera.

5.4. Keratome

The keratome has a sharp tip and both cutting edges. It is often used to enter the anterior chamber.

5.5. Blunt Tip Keratome

It is used for extension of the tunnel incision in cataract surgery.

6. Dissectors

6.1. Paton's Corneal Dissector

It is a delicate instrument used for dissection of cornea during lamellar keratoplasty.

6.2. Dastoor's Lacrimal Sac Dissector (Blunt)

It is a blunt dissector used for the dissection of lacrimal sac during dacryocystorhinostomy (DCR) operation as well as in other sac surgeries. Since it is a blunt instrument, the injury to the soft tissue structure is minimal.

6.3. Lang's Lacrimal Sac Dissector (Sharp)

It is similar to Dastoor's lacrimal dissector except for the sharp end.

7. Forceps

7.1. Fixation Forceps

The fixation forceps may have narrow or wide jaws. It may be with or without locking device. Usually, there are 2 × 3 teeth at the tips of the forceps; the number may vary from 1 × 2 to 4 × 5. It is used to fix the eyeball during surgery.

7.2. Dastoor's Superior Rectus Forceps

The superior rectus forceps has a S-shaped tip with 1 × 2 teeth. It is a strong forceps used to hold the superior rectus muscle in order to pass a bridle suture to fix the eyeball in downward gaze.

7.3. Moorfield's Forceps

The plain pointed Moorfield's forceps possesses a groove parallel to the limbs. It is used to hold the conjunctiva and sutures during surgery.

7.4. Wills Hospital Utility Forceps

Utility forceps is like Moorfield forceps but it has criss-cross serrated tips. It is used to hold the conjunctiva or skin in blunt dissection.

7.5. McPherson Corneal Forceps

The forceps has fine limbs having 1 × 2 teeth at the tip and a tying platform. It is used to hold the cornea while passing sclerocorneal sutures and to retract the cornea during lens delivery by cryoprobe.

7.6. St. Martin Forceps

It is a straight forceps with a long tying platform and 1 × 2 teeth at the tip. It is used for holding the cornea and the sclera and for suture tying.

7.7. Barraquer Colibri Forceps

It is a delicate forceps with a tying platform and 1 × 2 teeth. It is used for holding the corneal or the scleral flap as well as for suture tying.

7.8. Kirby's Iris Forceps

The forceps is small and delicate with fine limbs having 1 × 2 teeth on the inner side of the limbs. It is used to hold the iris while doing iridectomy during glaucoma and cataract surgeries or for optical purpose.

7.9. Bishop-Harmon Forceps

It is a straight tissue forceps with 1× 2 teeth. It is mainly used to hold the iris for doing iridectomy.

7.10. Lim's Forceps

It has a curved shaft with 1 × 2 teeth on the tip and a tying platform. It is used for holding the cornea and the sclera and for tying the suture.

7.11. Utrata's Capsulorhexis Forceps

In Utrata's forceps the tips are bent downward. The tips have flat platforms to hold the capsule to perform capsulorhexis.

7.12. Intracapsular Forceps

Two types of intracapsular forceps, Arruga's and Elschnig's, are popularly employed for intracapsular cataract extraction. Arruga's intracapsular forceps has a cup on inner side of the tip of each limb. The margins of the cups are smooth and on closure they do not bite the lens capsule. Elschnig's forceps has a double curve ending on a blunt point.

Arruga

Elsching

7.13. Intraocular Lens Insertion Forceps

Two types of intraocular lens (IOL) insertion forceps are commonly used—Lieberman's and Kelman-McPherson's. Lieberman's forceps has a ring at the tip for getting good grasp of an IOL. Kelman-McPherson's forceps has an angled platform which is ideal for holding and implanting the lens.

7.14. Lens Folding Forceps

It is designed to fold soft IOLs for insertion through small incisions.

7.15. Jaffe's Suture Tying Forceps

It can be either a straight or a curved type. It has a long tying platform for holding fine monofilament nylon and prolene sutures.

7.16. Jewelers Forceps

It can be either straight or curved shaft forceps with fine pointed tips. It is used for suture removal.

7.17. Vitreous Forceps

It is a very delicate forceps used for removal of an intraocular foreign body from the vitreous cavity.

7.18. Hartman's Mosquito Forceps

It is scissor-shaped forceps with blunt tips. It may be curved or straight with multiple grooves at right angle to the axis. The forceps is used to catch the bleeding vessels and hold the sutures during surgery.

7.19. Jameson's Muscle Forceps

It is a forceps-shaped clamp having teeth on the end of one of its curved limbs. The advancement forceps has a locking device. It is used to hold the muscle during strabismus surgery.

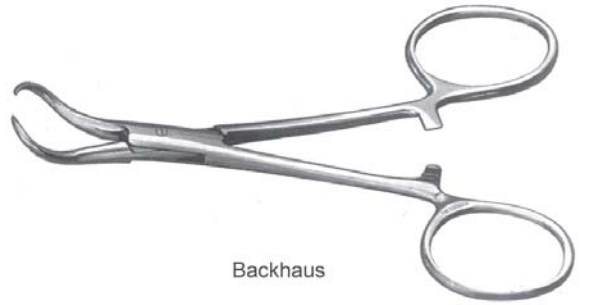
7.20. Epilation Forceps or Beer's Cilia Forceps

It is a small stout forceps with blunt flat ends. It is used to epilate the cilia in patients with trichiasis.

8. Clamps

8.1. Backhaus and Baby Jones towel clamps

The towel clamps are used to hold the sterilized drapes, tubings and cords during an ocular surgery.



Baby Jones

8.2. Berke's Ptosis Clamp

The clamp has J-shaped ends with internal serrations. It has a locking device. It is used to hold the levator muscle during ptosis surgery.

8.3. Snellen's Entropion Clamp

It consists of a D-shaped plate apposed by a U-shaped clamp. The clamp can be tightened with the help of a screw. It is a self-retaining instrument and provides good hemostasis. The metal plate not only protects the cornea but also supports the lid during surgery. It is used to correct entropion.

8.4. Lambert's Chalazion Clamp

It is a forceps having a screw for fixing the limbs like a clamp. One arm of the clamp has a round flat disc while the other arm has a circular ring of disc size. It is used to fix the chalazion during surgery and to obtain hemostasis by the pressure of the ring on the plate.

9. Scissors

9.1. McPherson-Westcott scissors

It is a curved small blade scissors with blunt tips. It is used to cut the conjunctiva.

9.2. Jaffe's Stitch Scissors

It has small blades with very sharp pointed tips and is used for removal of sutures.

9.3. Westcott's Tenotomy Scissors

It is a curved scissors having medium size blades with blunt tips. It is used for tenotomy in strabismus surgery.

9.4. Knapp's Strabismus Scissors

It is a plain straight or curved scissors with blunt ends. It is used to cut the extraocular muscles during strabismus surgery.

9.5. Castroviejo's Corneal Scissors

It is an angled scissors with blunt tipped small blades. It is used for keratoplasty and for making corneal or corneoscleral incision in extracapsular or intracapsular cataract extraction.

9.6. de Wecker's Scissors

It is a fine scissors with small blades directed at right angle to the arms. The blades are kept apart by the spring action of the arms. It is used for iridectomy.

9.7. Vannas Scissors

It is a fine delicate spring scissors having small straight or curved blades. It is used for cutting the anterior capsule of the lens during extracapsular cataract extraction (ECCE).

9.8. Vitreous Scissors

It is available in two styles, with straight and angled jaws. It is used in vitrectomy for cutting vitreous bands and membranes.

9.9. Enucleation Scissors

It is a stout scissors having curved blades with blunt ends. It is used for severing the optic nerve during enucleation.

10. Hooks, Expressors and Lens Manipulators

10.1. von Graefe's Muscle Hook

It has a round hook without any knob. It is used to separate the extraocular muscle from its scleral bed during strabismus surgery.

10.2. Jameson's Muscle Hook

The hook has a knob to engage and retain the extraocular muscle during strabismus surgery.

10.3. Chavasse's Muscle Hook

The hook has double curves to prevent the slippage of exposed muscle.

10.4. Sinskey's Lens Manipulating Hook

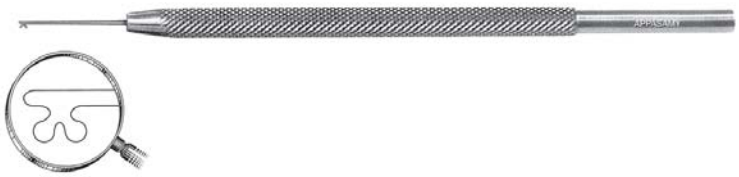
It is of two types, straight or angled, with a blunt tip. It is used to manipulate the lens during phacoemulsification.

10.5. Little's No-Hole Lens Manipulator

It is used to position the intraocular lens without any dialing hole.

10.6. Kuglen Hook

It is a small blunt hook with clove-shaped end. It is used for stretching the iris in nondilating pupil or manipulating the intraocular lens.



10.7. Agarwal's Chopper and Rotator

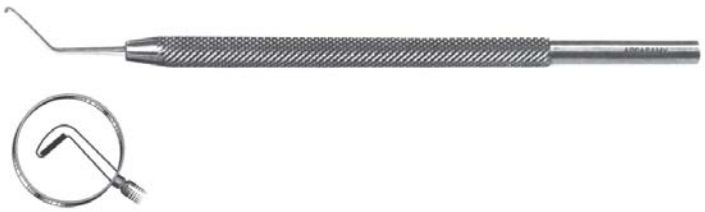
It has a cutting edge for chopping the lens and the 'Y'-shaped rotator to rotate the lens during phacoemulsification.

10.8. Kirby's Lens Expressor

One end of the instrument has a lens hook while the other has a flat spatula. The hook is used to express the lens during ECCE or ICCE. The spatula is used to reposit the iris.

10.9. Rosen's Phaco Splitter

It is an angled hook with sharp fine cutting edges. It is used for splitting (chopping) the nucleus during phacoemulsification.



11. Spatula, Spoon and Scoop

11.1. Dastoor's Iris Spatula

It is a delicate, flat, malleable spatula with blunt edges and tips. It is used to reposit the iris after iridectomy.

11.2. Castroviejo's Cyclodialysis Spatula

It is a flat blunt-edged spatula with blunt tip. To fit the convexity of the globe it is concave downwards and attached at right angle to the metallic handle. It is used to detach the ciliary body from the scleral spur during cyclodialysis operation for aphakic glaucoma.

11.3. Stallard's Lid or Ptosis Spatula

It is a metal plate having slightly convex surfaces with two holes at one end and six in the middle to pass sutures. It supports the lid and protects the cornea during ptosis surgery.

11.4. Jaeger's Lid Plate

It is a stout metallic plate with slight anterior convexity. It is used in ptosis surgery.

11.5. Mule's Evisceration Spoon

It has an oval or rectangular shallow cup attached to a stout metallic handle. It is used to scoop out the intraocular contents during evisceration.

11.6. Wells' Enucleation Spoon (Guide)

It is a spoon-shaped instrument with a central cleavage. It is used to engage the optic nerve during enucleation.

11.7. Meyerhoefer's Chalazion Scoop

It has a small cup with sharp edges attached to a narrow handle. It is used to scoop out the contents of a chalazion.

12. Needles, Canulas and Cystotome

12.1. Atkinson's Needle

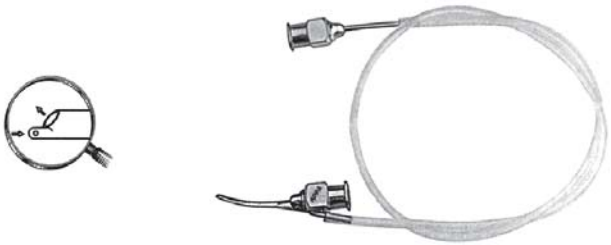
It is a long needle (23, 24 or 25 gauge, 1 or 1½ inches in length) used for peribulbar or retrobulbar anesthesia.

12.2. Short Needle

Short needles (26, 27 or 30 gauge, ½ inch in length) are used for subconjunctival, sub-Tenon or intravitreal injections.

12.3. Simcoe Irrigation-Aspiration Canula

It is a two-way metal canula having a silicone-tubing. The aspiration port always faces upwards. The conventional canula has the irrigation tip on the right side, while in the reverse type it is on the left. It is used for removing the cortical matter during ECCE and washing blood and viscoelastics from the anterior chamber.

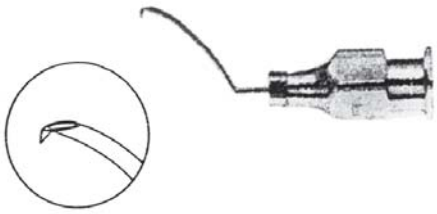


12.4. Rycroft's Anterior Chamber Canula

It is a curved canula with an angulated tip. It is used in ECCE for hydro procedures as well as in any intraocular surgery for injecting air/Ringer's lactate or balanced salt solution in the anterior chamber.

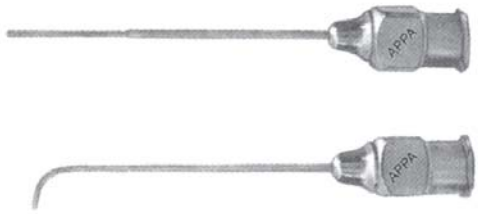
12.5. Blumenthal's Canula

It is 27 gauge needle with the shaft and the tip bent to 90°. It is used as an irrigating cystotome in small incision cataract surgery.



12.6. Lacrimal Canula

Lacrimal canulas are available in two styles: straight and curved. These are used in syringing of the lacrimal passage.

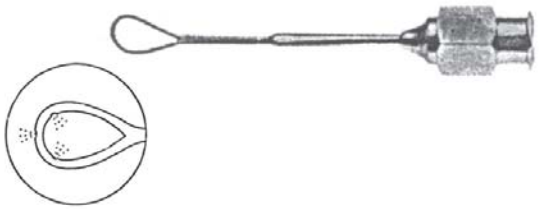


12.7. Cystotome

It is a tiny bent needle-knife, sharp on both the edges. It can be indigenously made by bending a 26 or 27 gauge disposable needle at its hub and bevel. It is used for doing anterior capsulotomy or capsulorhexis during ECCE, and posterior capsulotomy in posterior capsule opacification.

13. Drews' Irrigating Vectis

It consists of an oval loop measuring 6 mm wide and 9 mm long with one forward and two reverse irrigating ports. It is used in SICS and ECCE.



14. Needle Holders

14.1. Arruga's Needle Holder

It is a stout needle holder with a locking device. It is used to hold the needle during suturing and also for passing the bridle suture in the superior rectus muscle.

14.2. Kalt's Needle Holder

It is quite similar to Arruga's needle holder. It is used to hold the needle in lid, lacrimal and strabismus surgeries and also for passing the bridle sutures in rectus muscles.

14.3. McPherson's Needle Holder

The needle holder has a spring handle with finely serrated jaws to hold the needle.
It is used for microsurgery of the eye.

14.4. Troutman's Needle Holder

It is a delicate needle holder with curved jaws and is available with lock or without lock. It is used for passing fine monofilament sutures.

15. Barraquer's Blade Breaker

It has flat jaws and a locking device. It is used to hold the razor-blade fragment for making the corneoscleral groove and entering the anterior chamber.

16. Trephines and Punches

16.1. Elliot's Scleral Trephine

The trephine is available in three sizes: 1 mm, 1.5 mm and 2.0 mm. The trephine fits in a corrugated metal handle. It was used earlier for trephine operation for glaucoma.

16.2. Castroviejo's Corneal Trephine

Different diameters (5 mm to 11 mm) corneal trephines with adjustable stops are used for lamellar or penetrating keratoplasty.

16.3. Holth's Corneoscleral Punch

It is like a corneal scissors with a solid sharp thick blade which presses into a hollow rectangular frame thus cutting a piece of sclera. It is used to perform sclerectomy in glaucoma surgery.

16.4. Kelly's Punch

It is a Descemet's punch with spring action. It is used in filtration surgery to cut the trabecular meshwork.

17. Calipers and Markers

17.1. Castroviejo's Calipers

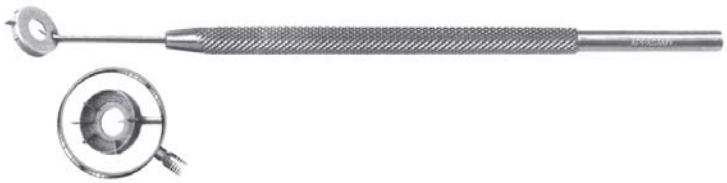
It is a divider-like instrument in which a graduated scale is attached to one arm, while the other arm can be moved by a screw over the scale. It is used to obtain measurements during strabismus, ptosis and retinal detachment surgeries.

17.2. Osher's Incision Calipers

It consists of two arms with a caliper. It is used for measurement of cataract incision especially in phacoemulsification and small incision cataract surgery.

17.3. LASIK Marker

It is a disk-shaped instrument with a central pin. It is used to mark the flap area for LASIK surgery.



17.4. Thornton's Optical Zone Marker

The diameter of the optical zone marker varies from 3 to 5 mm. It is used in refractive surgery to mark the optical zone.

18. Kansas' Lens Loop

It is a serrated loop attached to a stout handle used for removal of the nucleus or nuclear fragments in SICS.

19. Nightingale's Capsule Polisher

Nightingale's capsule polisher has a ring-shaped tip with semi-sharp edges. It is used for polishing the posterior capsule in ECCE and phacoemulsification.

20. Shepard's Fixation Ring

It is a C-shaped fixation ring with atraumatic teeth and swivel handle. It is used for fixing the eyeball while making a tunnel incision.

21. Dastoor's Keratoplasty Spatula

It is a delicate, angled, spoon-shaped spatula used in keratoplasty.

22. Tudor-Thomas Corneal Graft Stand

It is a metal stand used for preparation of the donor corneal graft.

23. Harm's Trabeculotome

It has a handle with two blunt upper and lower prongs. It is used in trabeculotomy, an operation for congenital glaucoma.

24. McPherson's Curved Bipolar Forceps

It is a bipolar forceps used for cauterization of bleeding points during ocular surgery.

25. Ball-point Cautery

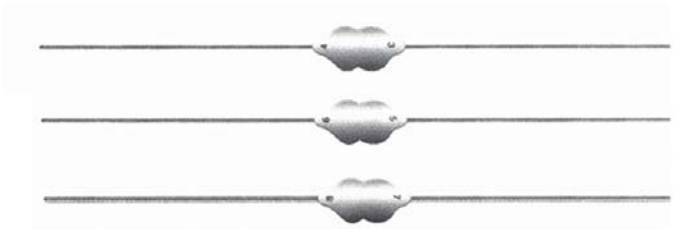
It has a round metal ball for retaining the heat and blunt tapering end for cauterization. It is a heat cautery used to cauterize bleeding vessels, especially after fashioning the conjunctival flap in cataract surgery and glaucoma filtration surgery.

26. Nettleship's Punctum and Canaliculus Dilator

It has a corrugated cylindrical metal handle with conical pointed tip. It is used to dilate the punctum and canaliculus before syringing or probing.

27. Bowman's Lacrimal Probes

These are a set of straight or curved metal wires of varied thickness (size 0 to 8) with bulbous ends. These are used to probe the nasolacrimal duct.



28. West's Lacrimal Chisel

It has a flat sharp tapering blade attached to a stout metallic handle. It is used to cut the bones during DCR operation.

29. Mallet's Hammer

It is a small steel hammer attached to a corrugated handle. It is used to hammer the chisel during DCR operation.

30. Traquair's Periosteal Elevator

It has two ends, one end is blunt while the other is semi-sharp. It is used in DCR operation and orbitotomies.

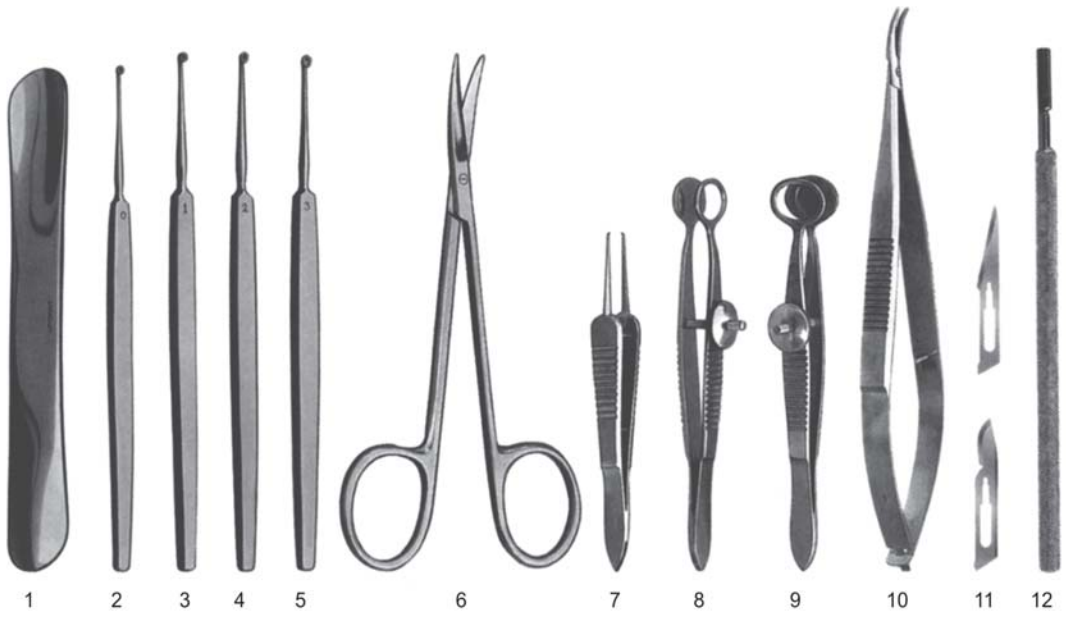
31. Citeli Bone Nibbler or Bone Punch

The punch consists of a spring handle and two blades. The upper blade has a hole with cutting edges and the lower blade has a cup-like depression. It is used to cut the bone in DCR operation.

32. Complete Surgical Sets

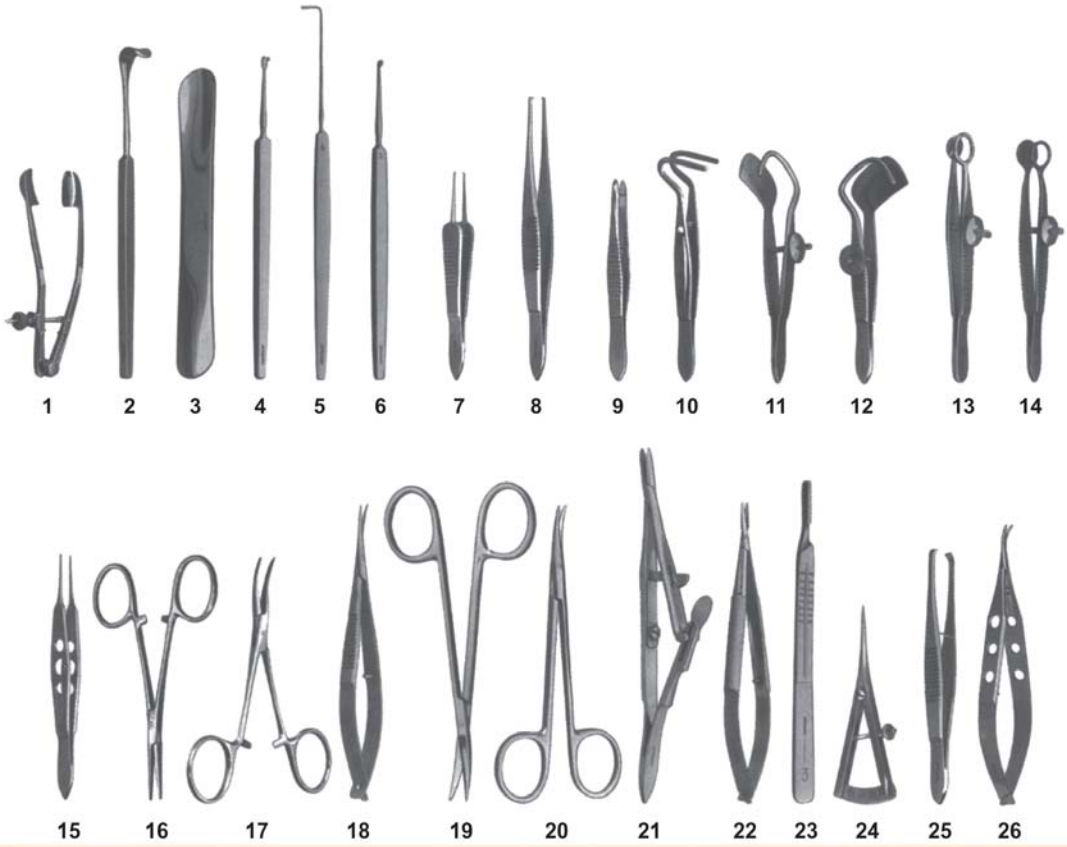
The names and figures of various instrument-sets used in different ocular surgical procedures are given below:

32.1. Instruments used for Chalazion Surgery



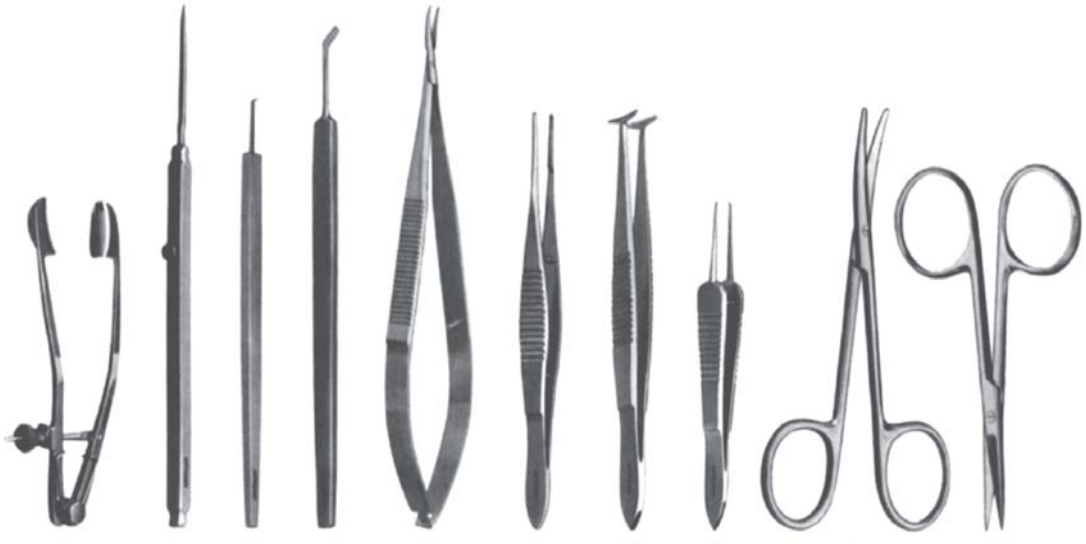
1	Jaeger lid plate
2	Meyerhoefer chalazion curette, size 0, 1.50 mm
3	Meyerhoefer chalazion curette, size 1, 1.75 mm
4	Meyerhoefer chalazion curette, size 2, 2.25 mm
5	Meyerhoefer chalazion curette, size 3, 2.50 mm
6	Eye scissors, Curved
7	St. Martin forceps, 1x2 Teeth
8	Lambert chalazion clamp, 12 mm
9	Desmarres chalazion clamp, 13 mm/20 mm
10	Castroviejo needle holder
11	Bard-Parker blade # 11 and # 15
12	Bard-Parker handle

32.2. Instruments Used for Lid Surgery



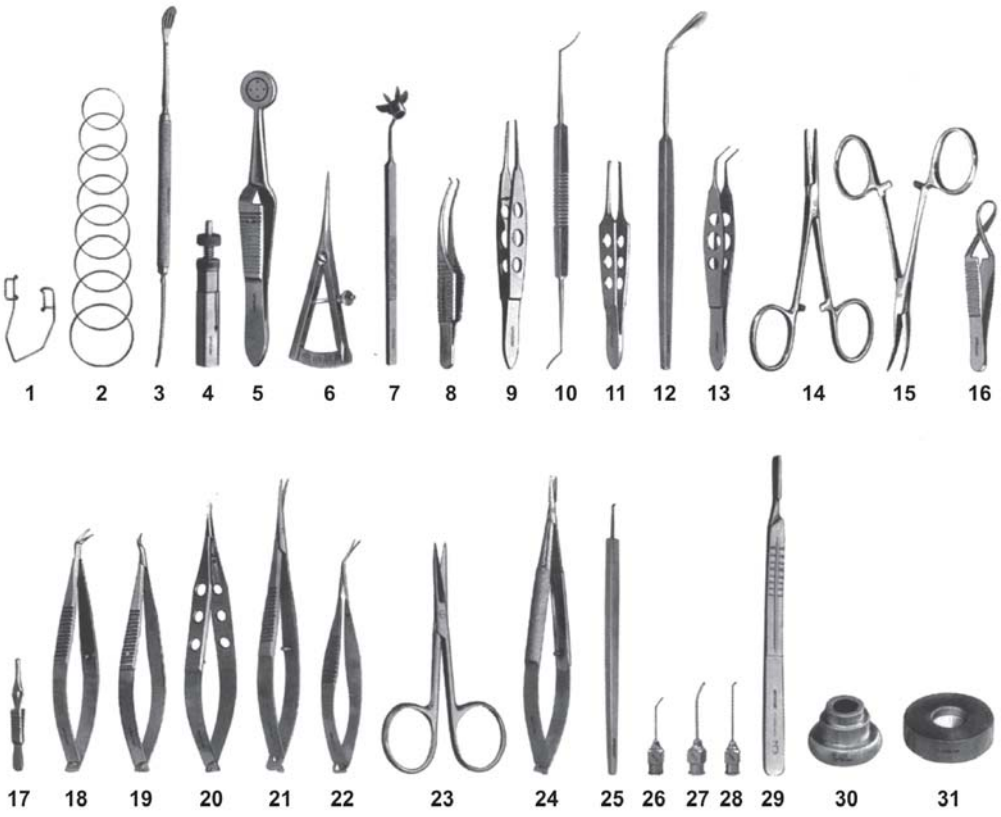
1	Lancaster eye speculum	14	Lambert chalazion clamp
2	Desmarres lid retractor	15	McPherson tying forceps
3	Jaeger lid plate	16	Hartman mosquito forceps, straight
4	Fixation hook	17	Hartman mosquito forceps, curved
5	Graefe muscle hook	18	Westcott stitch scissors
6	Meyerhoefer chalazion curette	19	Eye scissors, curved
7	St. Martin forceps	20	Stevens tenotomy scissors, curved
8	Fixation forceps, 1x2 Teeth	21	Kalt needle holder
9	Beer cilia forceps	22	Barraquer needle holder
10	Berke ptosis clamp	23	Bard - Parker handle
11	Snellen entropion forceps	24	Castroviejo caliper
12	Snellen entropion forceps	25	Fixation foreceps, 2x3 teeth
13	Ayer chalazion clamp	26	Corneal scissors

32.3. Instruments Used for Pterygium Surgery



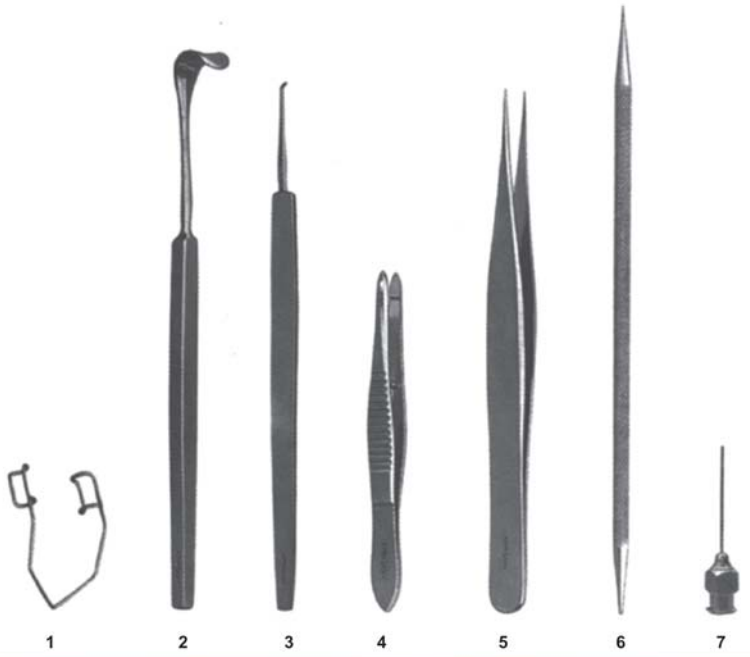
1 Lancaster eye speculum
2 Barraquer cataract knife
3 Paufique graft knife
4 Paton corneal dissector
5 Castroviejo needle holder
6 Bonaccolot utility forceps
7 Green fixation forceps
8 St. Martin suturing forceps
9 Strabismus scissors, curved
10 Iris scissors, straight

32.4. Instruments Used for Corneal Transplantation



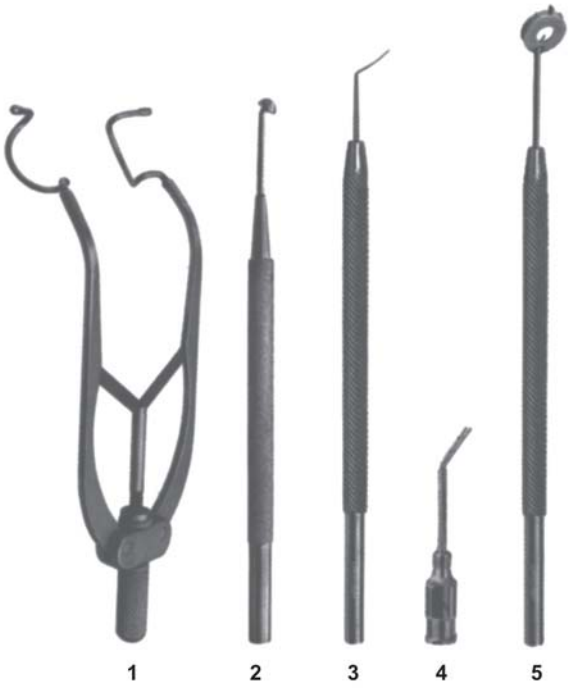
1 Barraquer wire speculum
2 Flieringa scleral fixation ring (set of 8 sizes)
3 Paton spatula and spoon
4 Castroviejo corneal trephine
5 Dastoor corneal graft holding forceps
6 Castroviejo caliper
7 Osher-Neumann radial marker, 8 blades
8 Colibri forceps, 1x2 teeth
9 McPherson corneal forceps, 1x2 teeth
10 Castroviejo cyclodialysis spatula
11 Bishop-Harmon tissue forceps
12 Dastoor keratoplasty spatula
13 Kelman-McPherson forceps
14 Hartman mosquito forceps, straight
15 Hartman mosquito forceps, curved
16 Baby Jones towel clamp
17 Serrefine clamp
18 Castroviejo corneoscleral scissors, left
19 Castroviejo corneoscleral scissors, right
20 Castroviejo corneoscleral scissors, universal
21 Westcott stitch scissors
22 Vannas capsulotomy scissors
23 Iris scissors
24 Barraquer needle holder
25 Paufique graft knife
26 Rycroft air injection canula, 27 G
27 Bracken anterior chamber canula, curved, 19 G
28 Knolle anterior chamber Irrigating canula, 23 G
29 Bard-Parker handle
30 Tudor-Thomas corneal graft stand
31 Lieberman teflon block

32.5. Instruments Used for Removal of Superficially Lodged Foreign Body



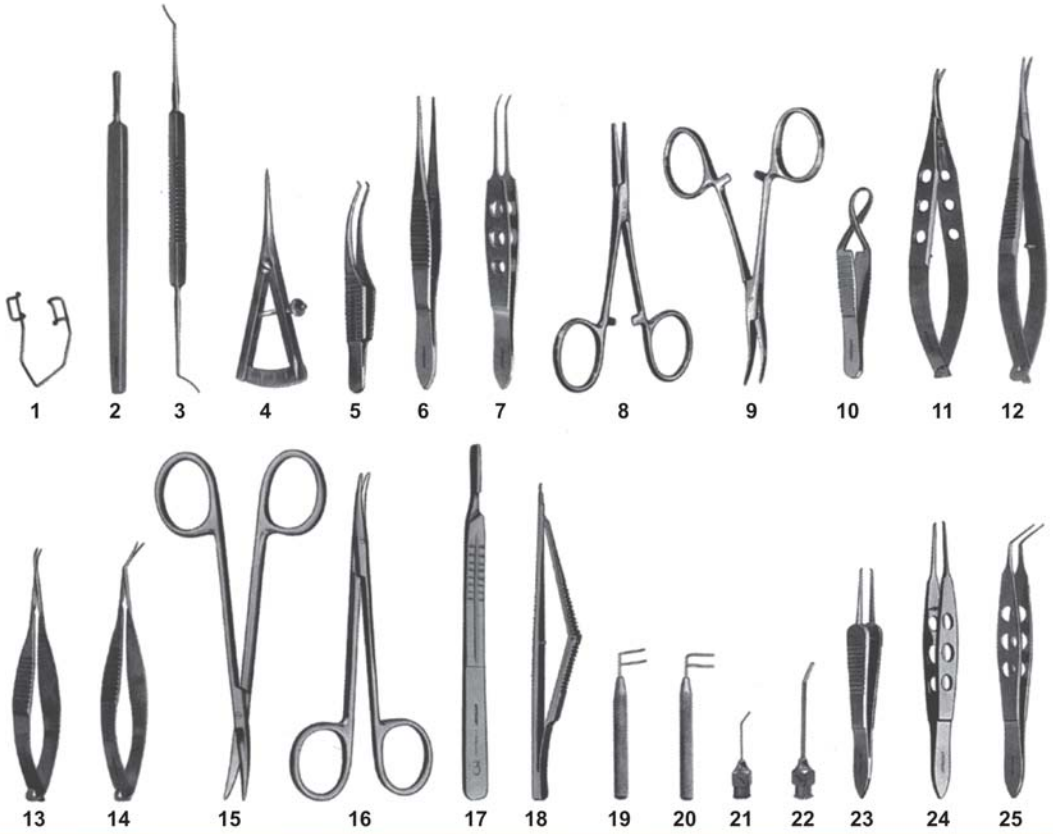
1 Barraquer wire speculum
2 Desmarres lid retractor
3 Golf club foreign body spud
4 Beer cilia forceps
5 Jewelers forceps
6 Castroviejo lacrimal dilator
7 Lacrimal canula, straight, 23 G

32.6. Instruments Used for LASIK Surgery



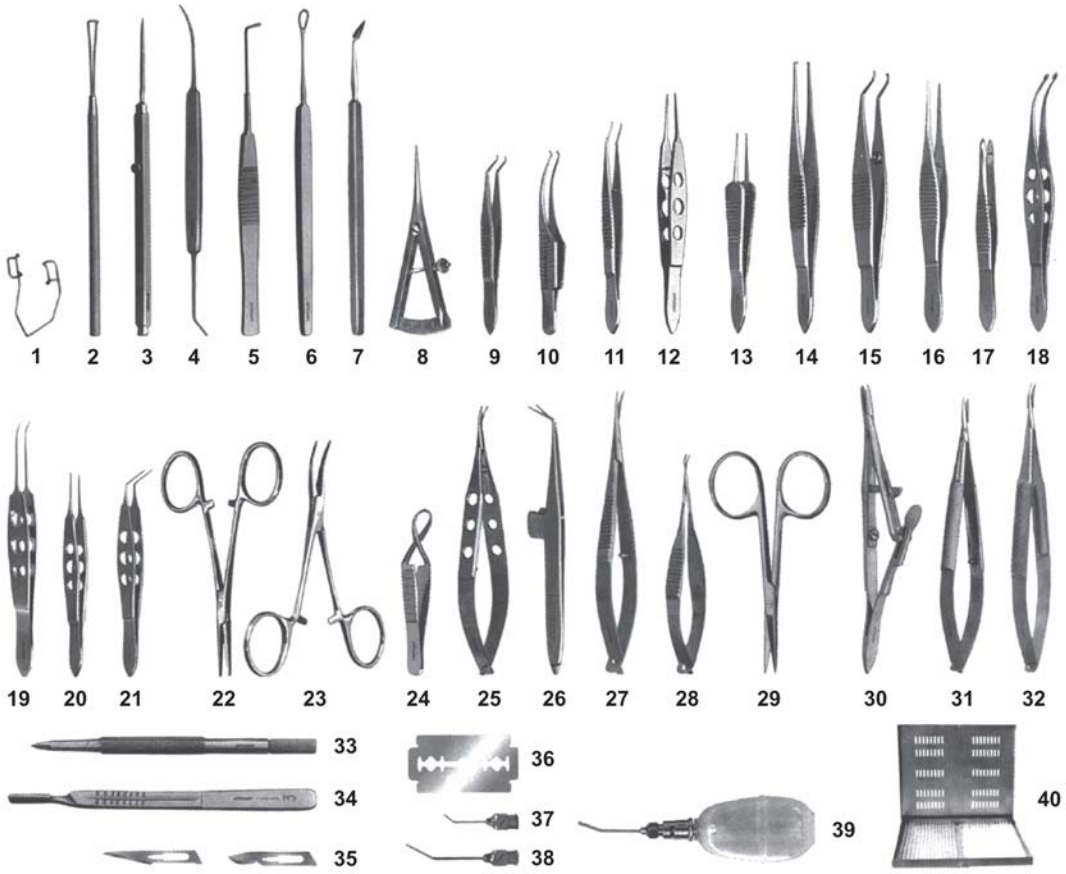
1 Castroviejo LASIK speculum
2 LASIK depressor
3 LASIK spatula
4 LASIK flap irrigator
5 LASIK marker

32.7. Instruments Used for Glaucoma Surgery



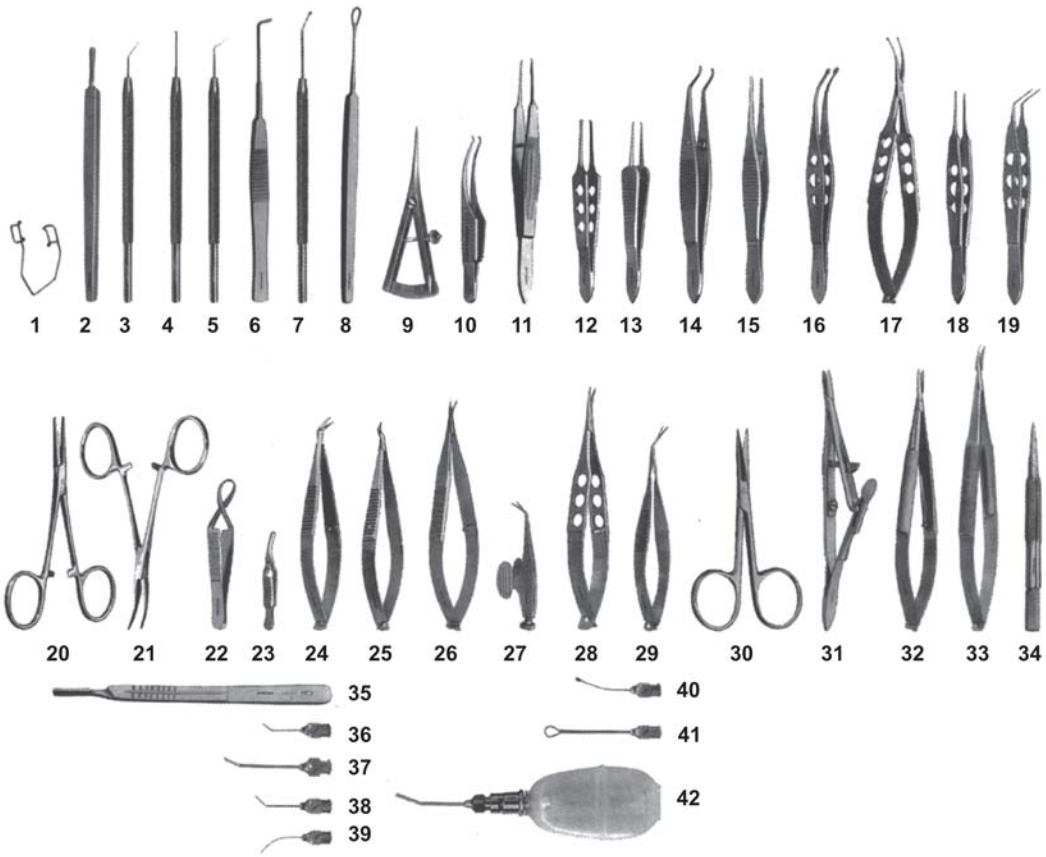
1 Barraquer wire speculum
2 Tooke corneal knife
3 Iris spatula
4 Castroviejo caliper
5 Colibri forceps, 1x2 teeth
6 Eye dressing forceps
7 Jaffe tying forceps, curved
8 Hartman mosquito forceps, straight
9 Hartman mosquito forceps, curved
10 Baby Jones towel clamp
11 Castroviejo corneal scissors
12 Westcott stitch scissors
13 Vannas scissors, curved
14 Vannas capsulotomy scissors
15 Eye scissors, curved
16 Stevens tenotomy scissors, curved
17 Bard-Parker handle
18 Kelly Descement's punch
19 Harms trabeculotomy probe, right
20 Harms trabeculotomy probe, left
21 Rycroft air injection canula, 27 G
22 Anterior chamber washout canula, 16 G
23 St. Martin forceps
24 McPherson corneal forceps, 1x2 teeth
25 McPherson tying forceps, angled

32.8. Instruments Used for Intracapsular Cataract Extraction



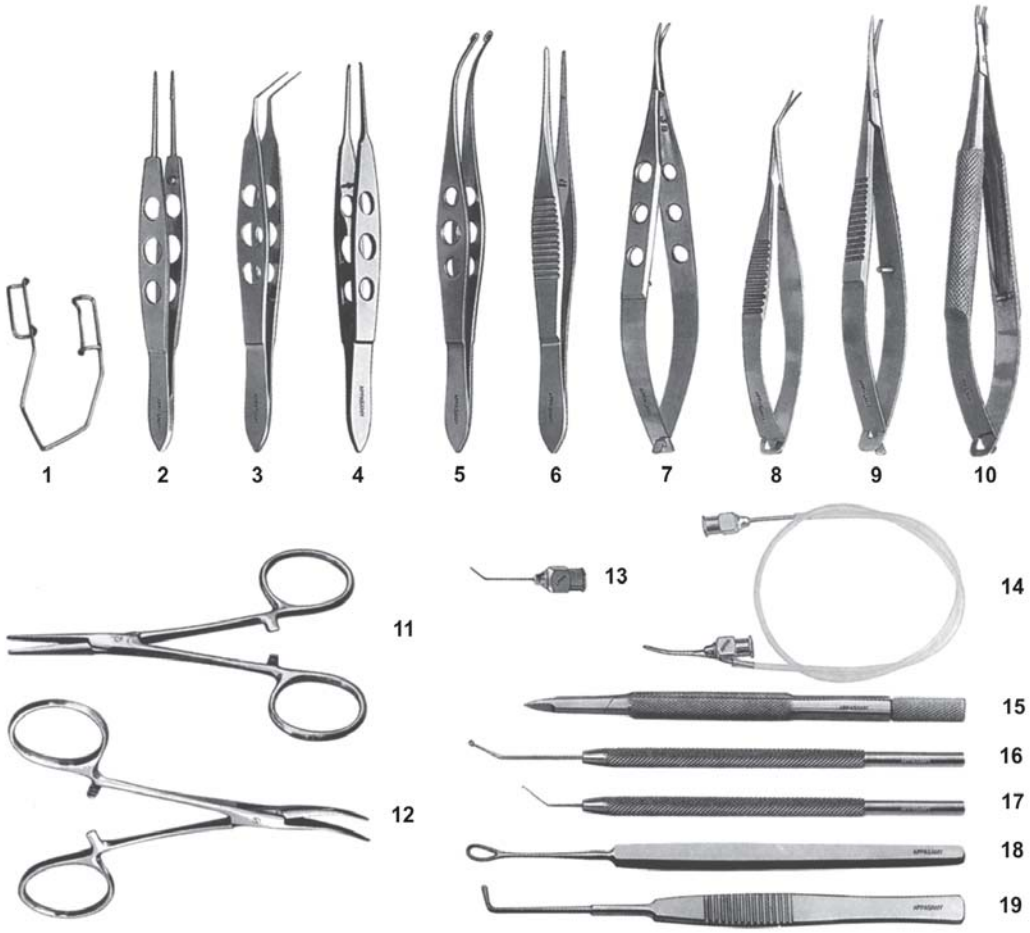
1	Barraquer wire speculum	21	McPherson forceps, angled
2	Dastoor iris retractor	22	Hartman mosquito forceps, straight
3	Barraquer cataract knife in sliding case	23	Hartman mosquito forceps, curved
4	Iris repositor	24	Baby Jones towel champ
5	Smith lens expressor	25	Castroviejo corneal scissors
6	Lewis lens loop	26	deWecker's iris scissors
7	Cautery, cone type	27	Westcott stitch scissors
8	Castroviejo caliper	28	Vannas capsulotomy scissors
9	Hess iris forceps, 1x2 teeth	29	Iris scissors
10	Colibri forceps, 1x2 teeth	30	Kalt needle holder
11	Lim's corneoscleral forceps 1x2 teeth	31	Barraquer needle holder
12	Mcpherson corneal forceps, 1x2 teeth	32	Barraquer needle holder, curved
13	St. Martin forceps	33	Castroviejo blade breaker and holder
14	Fixation forceps, 1x2 teeth	34	Bard-Parker handle
15	Dastoor superior rectus forceps, 1x2 teeth	35	Bard-Parker blade
16	Wills hospital utility forceps	36	Carbon razor breakable blade
17	Beer cilia forceps	37	Rycroft air injection canula, 23 G
18	Arruga capsule forceps	38	Bishop-Harmon anterior chamber canula, 19 G
19	Jaffe tying forceps, curved	39	Silicone blub with adaptor
20	McPherson tying forceps	40	Sterilization tray

32.9. Instruments Used for Extracapsular Cataract Surgery



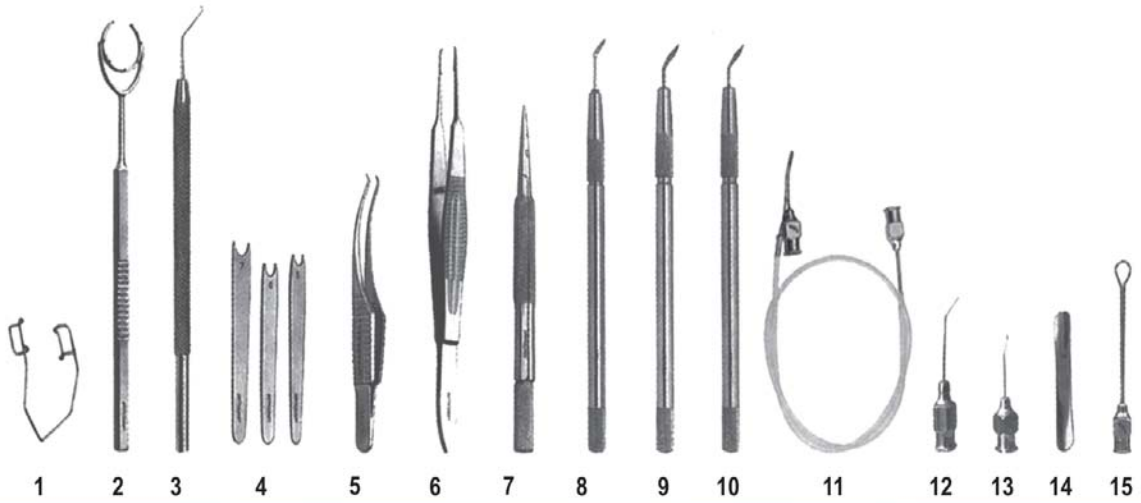
1	Barraquer wire speculum
2	Tooke corneal knife
3	Sinskey lens manipulating hook
4	Kuglen iris hook
5	Barraquer iris spatula
6	Smith lens expressor
7	Nightingale capsule polisher
8	Lewis lens loop
9	Castroviejo caliper
10	Colibri forceps, 1x2 teeth
11	Castroviejo suturing forceps
12	Bishop-Harmon tissue forceps
13	St. Martin suturing forceps, 1x2 teeth
14	Dastoor superior rectus forceps
15	Wills hospital utility forceps
16	Arruga capsule forceps
17	Shepard IOL forceps
18	McPherson tying forceps
19	McPherson forceps
20	Hartman mosquito forceps, straight
21	Hartman mosquito forceps, curved
22	Baby Jones towel clamp
23	Serrefine clamp
24	Castroviejo corneoscleral scissors, left
25	Castroviejo corneoscleral scissors, right
26	Micro iris scissors
27	Barraquer iris scissors
28	Westcott tenotomy scissors
29	Vannas capsulotomy scissors
30	Iris scissors, straight
31	Kalt needle holder, straight
32	Barraquer needle holder,
33	Barraquer needle holder curved
34	Swiss model blade breaker and holder
35	Bard Parker handle
36	Rycroft air injection canula, 16 G
37	Anterior chamber washout canula, 16 G
38	Pearce hydrosdissection canula
39	Kellsan hydrodelineation canula
40	Jensen capsule polisher
41	Knolle-Pearce irrigating vectis
42	Silicone bulb with adaptor

32.10. Instruments Used for Intraocular Lens Implantation



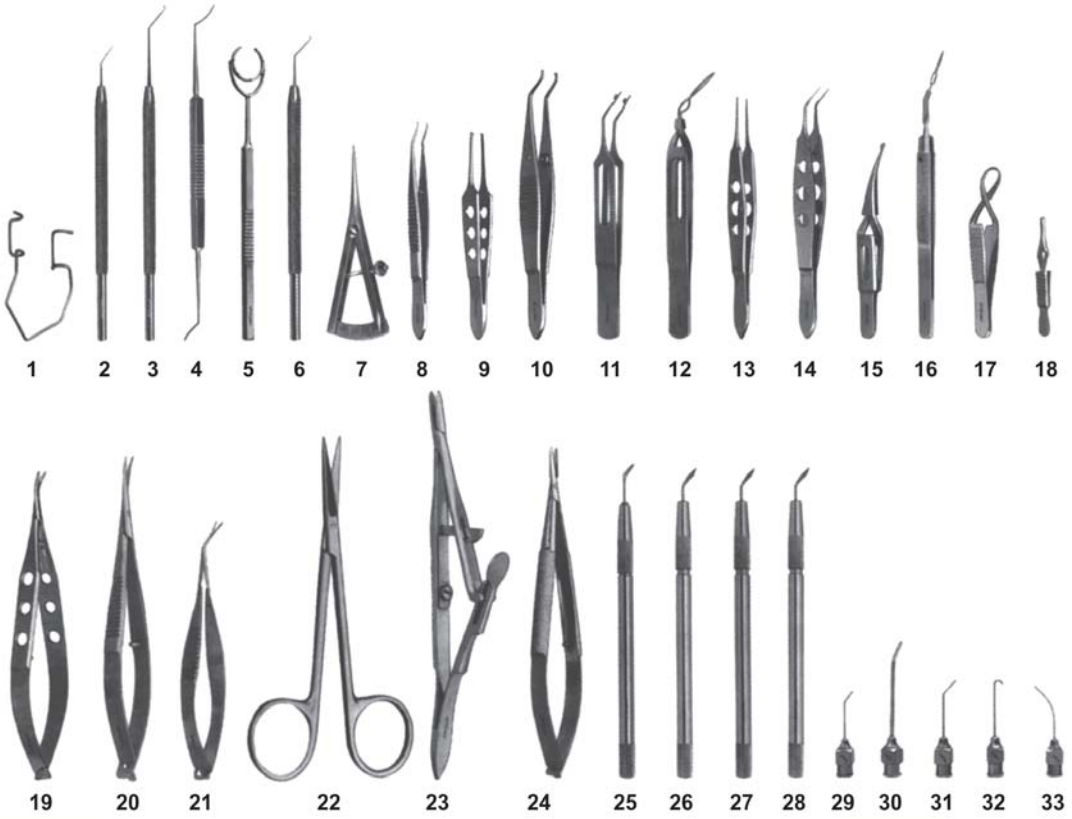
1 Barraquer wire speculum
2 Jaffe tying forceps
3 McPherson forceps, angled
4 McPherson corneal forceps, 1x2 teeth
5 Arruga capsule forceps
6 Wills hospital utility forceps
7 Castroviejo corneal scissors, universal
8 Vannas capsulotomy scissors, angled
9 Westcott stitch scissors
10 Barraquer needle holder
11 Hartman mosquito forceps, straight
12 Hartman mosquito forceps, curved
13 Rycroft air injection canula
14 Simcoe I/A canula
15 Castroviejo blade breaker
16 Nightingale capsule polisher
17 Sinskey lens manipulating hook
18 Lewis lens loop
19 Smith lens expressor

32.11. Instruments Used for Small Incision Cataract Surgery



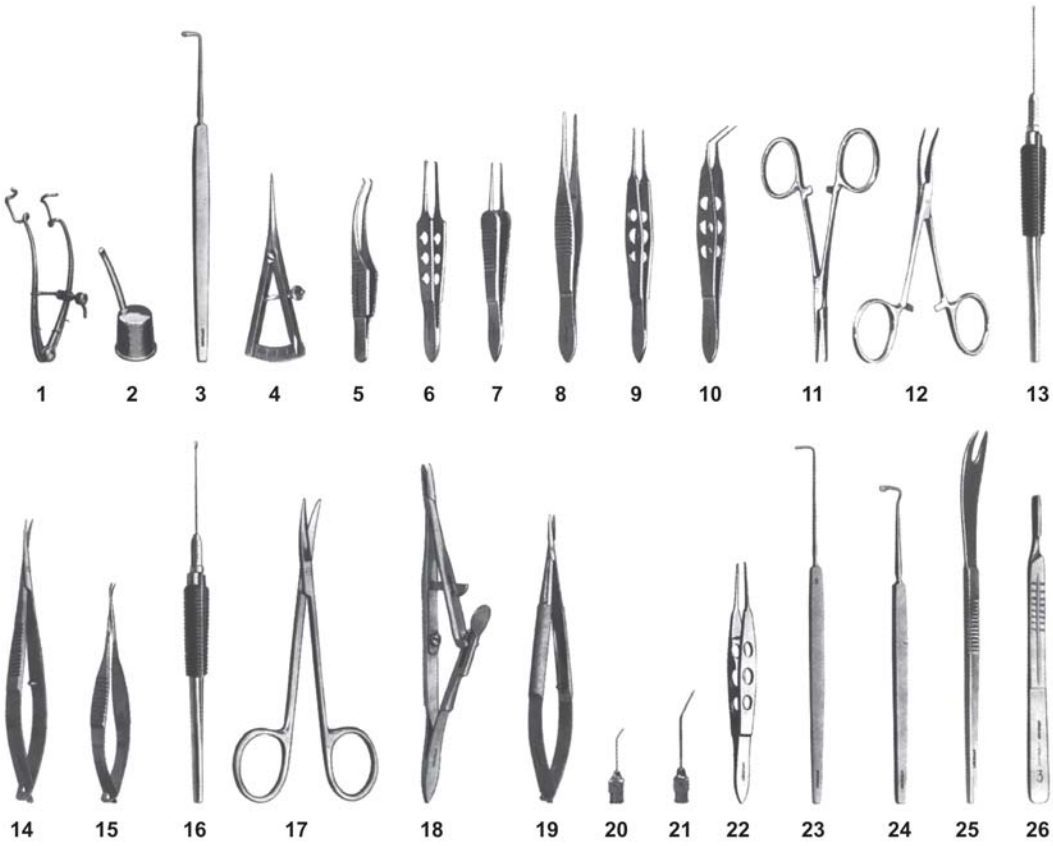
1 Barraquer wire speculum
2 Shepard fixation ring
3 Sinskey's hook
4 Galand incision marker
5 Colibri forceps
6 Castroviejo suturing forceps
7 Swiss model blade breaker and holder
8 Tunnel blade
9 Keratome
10 Extension blade
11 Simcoe canula
12 Anterior chamber canula, 25 G
13 Anterior chamber canula, 27 G
14 IOL glide
15 Irrigating vectis

32.12. Instruments Used for Phacoemulsification



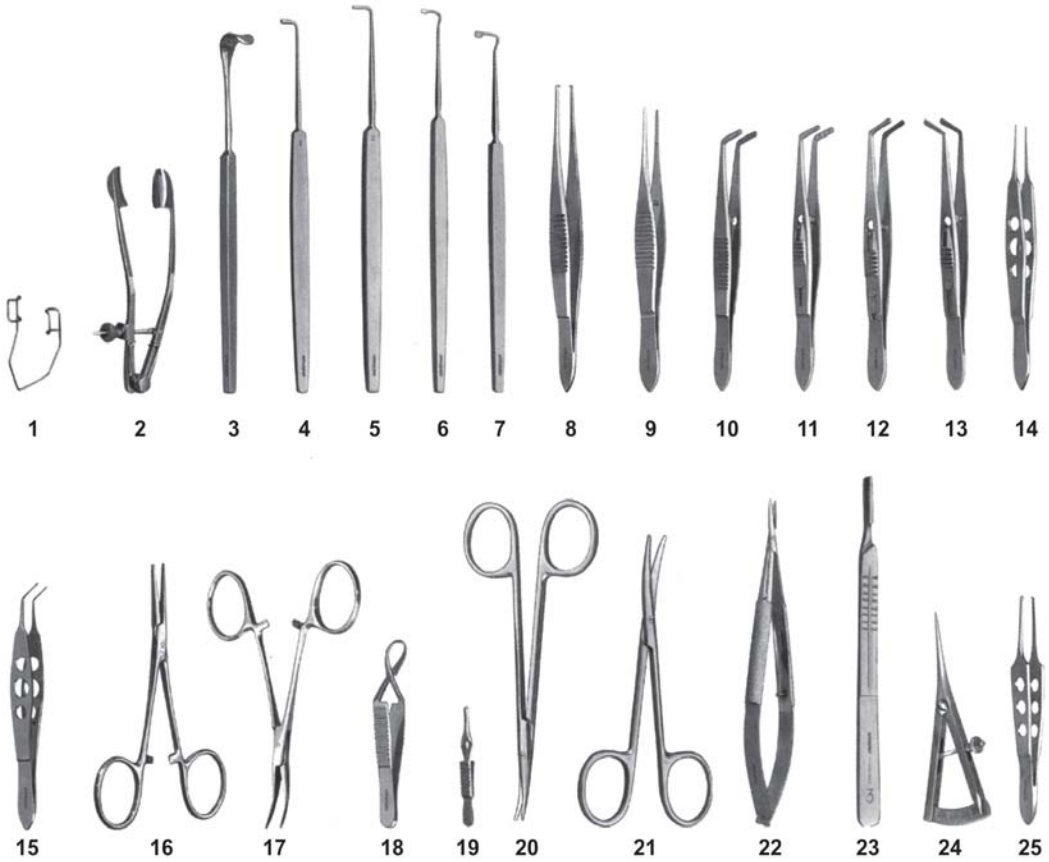
1 Kratz Barraquer wire speculum
2 Sinskey lens manipulating hook
3 Akahoshi nucleus sustainer
4 Castroviejo cyclodialysis spatula
5 Shepard fixation ring
6 Agarwal's phaco chopper
7 Castroviejo caliper
8 Lim's corneoscleral forceps
9 Bishop-Harmon tissue forceps
10 Dastoor superior rectus forceps
11 Lens folder
12 Lens inserting forceps
13 McPherson tying forceps
14 Utrata capsulorhexis forceps
15 Dodick nucleus cracker
16 Akahoshi prechop forceps
17 Baby Jones towel clamp
18 Serrefine clamp
19 Castroviejo cornesl scissors
20 Westcott stitch scissors
21 Vannas capsulotomy scissors
22 Eye scissors
23 Kalt needle holder
24 Barraquer needle holder
25 Tunnel blade
26 Keratome, 2.8 mm
27 Keratome, 3.2 mm
28 Extension blade
29 Rycroft air injection canula, 23 G
30 Anterior chamber washout canula, 16 G
31 Pearce hydrodissection canula
32 Gimbel 'U' shaped hydrodissection canula
33 Kellan hydrodelineation canula

32.13. Instruments Used for Vitreoretinal Surgery

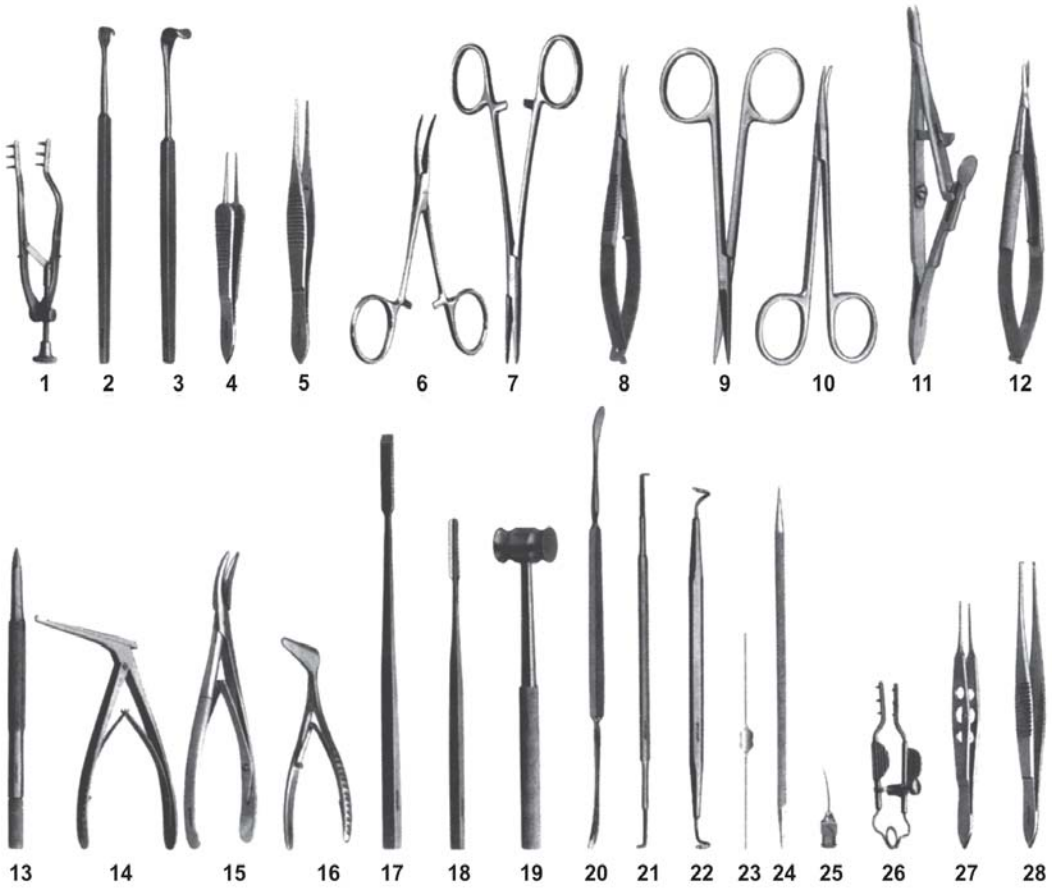


1 Clark eye speulum
2 Schepens scleral depressor
3 Gass retinal detachment hook
4 Castroviejo caliper
5 Colibri forceps, 1x2 teeth
6 Bishop-Harmon tissue forceps
7 St. Martin suturing forceps, 1x2 teeth
8 Wills hospital utility forceps
9 McPherson tying forceps
10 McPherson forceps
11 Hartman mosquito forceps, straight
12 Hartman mosquito forceps, curved
13 Vitreous forceps
14 Westcott stitch scissors
15 Vannas capsulotomy scissors
16 Vitreous scissors
17 Eye scissors, curved
18 Kalt needle holder
19 Barraquer needle holder
20 Rycroft air injection canula
21 Bishop-Harmon anterior chamber canula
22 McPherson corneal forceps
23 Graefe muscle hook
24 Jameson muscle hook
25 Schepens forked orbital retractor
26 Bard-Parker handle

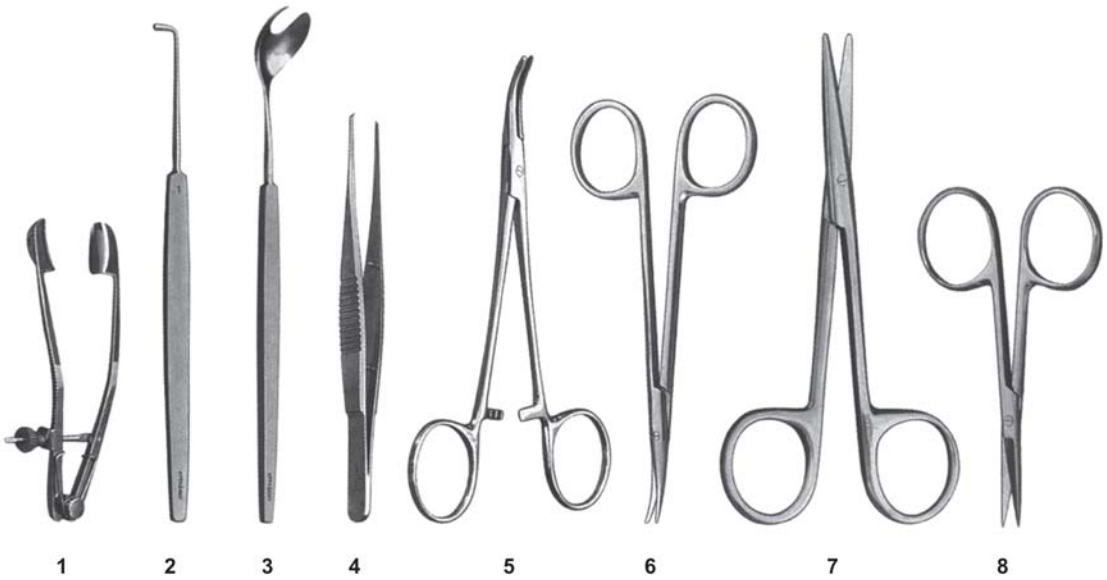
32.14. Instruments Used for Strabismus Surgery



1 Barraquer wire speculum
2 Lancaster eye speculum
3 Desmarres lid retractor
4 Graefe muscle hook, size 1
5 Graefe muscle hook, size 2
6 Jameson muscle hook, small
7 Jameson muscle hook, large
8 Fixation forceps, 1x2 teeth
9 Wills hospital utility forceps
10 Jameson muscle forceps, left
11 Jameson muscle forceps, right
12 Worth advancement forceps, left
13 Worth advancement forceps, right
14 McPherson tying forceps
15 Kelmen-McPherson forceps
16 Hartman mosquito forceps, straight
17 Hartman mosquito forceps, curved
18 Baby Jones towel clamp
19 Serrefine clamp
20 Stevens tenotomy scissors, curved
21 Knapp strabismus scissors, curved
22 Barraquer needle holder
23 Bard-Parker handle
24 Castroviejo caliper
25 Bishop-Harmon forceps



1 Stevenson lacrimal sac retractor
2 Knapp lacrimal sac retractor
3 Desmarres lid retractor
4 St. Martin suturing forceps, 1x2 teeth
5 Wills hospital utility forceps
6 Hartman mosquito forceps, curved
7 Halsted mosquito forceps, straight
8 Westcott stitch scissors
9 Eye scissors, straight
10 Stevens tenotomy scissors, curved
11 Kalt needle holder
12 Barraquer needle holder
13 Castroviejo blade breaker
14 Citeli bone nibbler
15 Beyer ronguer
16 Nasal speculum
17 West lacrimal chisel
18 West bone gauge
19 Mallet hammer
20 Freer periosteal elevator
21 Traquair periosteal elevator
22 Pigtail probe with suture holes
23 Bowman lacrimal probe
24 Castroviejo lacrimal dialator
25 Lacrimal canula, curved
26 Muller lacrimal sac retractor
27 McPherson tying forceps
28 Fixation forceps



1 Lancaster eye speculum
2 Graefe muscle hook
3 Wells enucleation spoon
4 Elschnig fixation forceps
5 Halsted mosquito forceps, curved
6 Stevens tenotomy scissors, curved
7 Enucleation scissors, straight
8 Eye scissors, straight

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D

E

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